

An Information Booklet

VASCULITIS



arc 0870 850 5000
www.arc.org.uk
Committed to curing arthritis

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Introduction

This booklet explains what vasculitis is, how it is recognized, how it is treated, and what you can do to help your treatment. All the main types of vasculitis are briefly explained and we also suggest other sources of information which may be helpful. Terms which appear in *italics* when they are first used are explained in the glossary at the back of the booklet. There are many different types of vasculitis. Although we do not know what causes many of these, there are treatments which can be very effective.

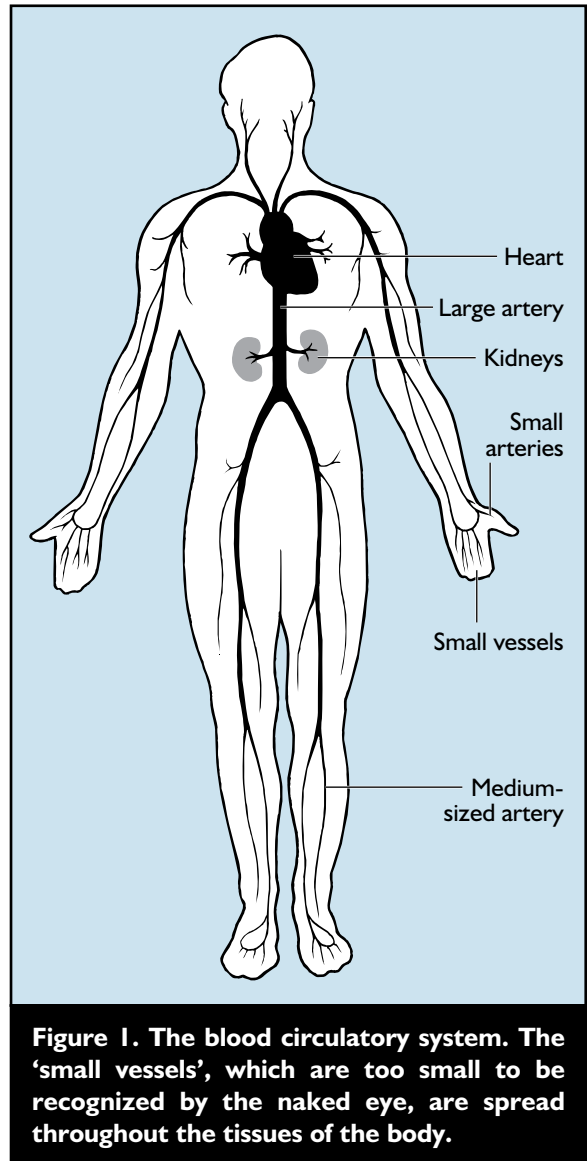
What is vasculitis?

Vasculitis means that the blood vessels are inflamed. When part of your body is inflamed, it swells and is usually uncomfortable or painful (although with many types of vasculitis you will not be able to see any swelling on the outside of the body). The term ‘-itis’ means inflammation, so, for example, when you have appendicitis, your appendix is inflamed; and when you have arthritis, your body joints are inflamed.

Blood vessels are the tubes which carry blood around your body. There are three types of blood vessel which can be affected by vasculitis:

- **arteries**, which take blood from the heart to various parts of the body – to organs such as the kidneys and liver, and to body tissues such as the skin
- **veins**, which take blood back to the heart
- **capillaries**, which are tiny vessels between the arteries and the veins where oxygen and other materials pass from the blood into the tissues.

Figure 1 shows where these blood vessels are in your body (the blood circulatory system).



Our body organs and tissues need a regular blood supply to work properly. If the blood vessels are inflamed, this can block or reduce the flow of blood, as shown in Figure 2. The wall of the blood vessel can also bulge – this is known as an aneurysm. An aneurysm can burst

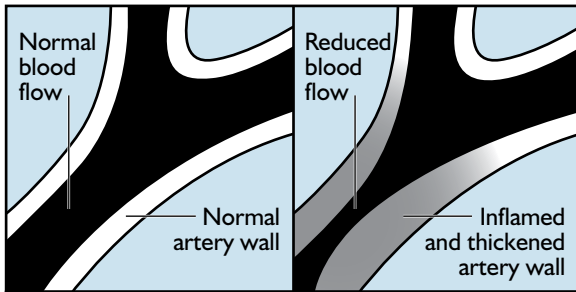


Figure 2. ‘Vasculitis’ means inflammation of the blood vessels. This diagram shows a normal artery (left) and one which is inflamed (right).

(rupture) and cause internal bleeding. The damage which vasculitis can cause depends upon which part of the body is affected – the bigger the blood vessel the greater the potential damage. And the more important the body tissue supplied, the more serious the damage will be.

What are the symptoms?

Vasculitis takes many different forms and the symptoms vary enormously from person to person. Many people with vasculitis feel unwell with fever, sweats, fatigue and weight loss. Other symptoms vary according to which part of the body is affected. Vasculitis in the skin causes a rash of spots which can sometimes rupture leaving open sores (ulcers). When vasculitis affects only the skin the long-term effects are not usually serious, and the symptoms generally clear up once the inflammation has settled. People with inflammation in their lungs may have a cough or be short of breath, while inflammation of the nerves can cause pins and needles or weakness in an arm or leg. When vasculitis affects the kidneys it can cause problems passing urine and there may also be blood in the urine. Unfortunately, the symptoms of kidney vascu-

litis often do not appear until the kidneys have been damaged and have started to function less effectively. If the damage is severe, treatment on an artificial kidney (*dialysis*) machine may sometimes be necessary.

Vasculitis can appear suddenly in someone who has previously been completely well – doctors call this primary vasculitis. Vasculitis can also appear in people who have an established disease such as arthritis – this is called secondary vasculitis. It may happen, for example, with rheumatoid arthritis, systemic lupus erythematosus (SLE) or Sjögren’s syndrome (see **arc** booklets ‘Rheumatoid Arthritis’, ‘Lupus’, ‘Sjögren’s Syndrome’).

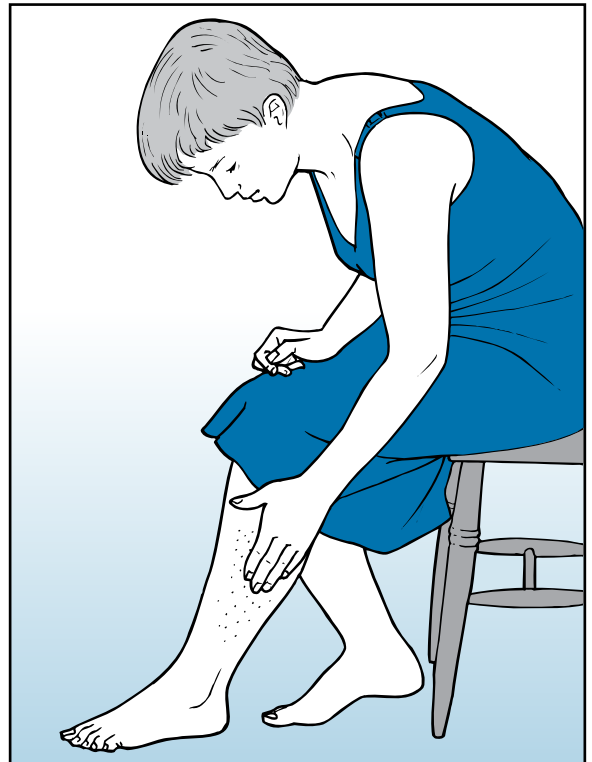


Figure 3. A rash on the legs is a common symptom with certain types of vasculitis.

Doctors usually define the types of vasculitis according to the size of the blood vessels involved. All these different types are explained later in this booklet:

- **Vasculitis in large arteries** – this includes temporal arteritis and Takayasu’s arteritis.
- **Vasculitis in medium-sized arteries** – this includes polyarteritis nodosa and Kawasaki disease. Medium-sized arteries can also be involved in vasculitis which occurs with (is secondary to) rheumatoid arthritis, systemic lupus erythematosus and Sjögren’s syndrome.
- **Vasculitis in small arteries** – this includes Wegener’s granulomatosis, microscopic polyangiitis and Churg–Strauss syndrome. Vasculitis in the small arteries can also be a consequence of rheumatic diseases, including rheumatoid arthritis and systemic lupus erythematosus. It can happen with infections including hepatitis and, very occasionally, with different types of cancers, including leukaemia and lymphomas.
- **Vasculitis in small vessels (usually capillaries)** – this usually involves the skin and is also sometimes caused by a reaction to certain drugs.

How common is vasculitis?

About 3,000 people in the UK develop vasculitis in any given year – about 5 people per 100,000 in the population (this is the number of new cases). Different types of vasculitis affect different age groups. Temporal arteritis, for example, which affects the arteries of the head, is much more common in people over 50, and it is fairly common for it to be associated with a condition called polymyalgia rheumatica (PMR) (see **arc** booklet ‘Polymyalgia Rheumatica (PMR)’). On the other hand, Henoch–Schönlein purpura (HSP), which is an allergic form of vasculitis mainly affecting the skin, is much more common in children than in adults.

What causes vasculitis?

There is no one cause, and in most cases the cause is unknown. We do know that vasculitis is not directly inherited – but genetic factors may play a part as several cases can occur in the same family. We also know what causes some types of vasculitis – for example, drugs and a variety of infections, particularly those associated with hepatitis, can cause small vessel vasculitis.

From the research done up to now, we believe that most forms of vasculitis are a type of autoimmune disease. This means that the body’s defence mechanisms are not doing their normal job of fighting infections and keeping us healthy. Instead, these mechanisms attack a healthy part of the body – the blood vessel wall. This causes the inflammation that damages the blood vessels.

What are the tests for vasculitis?

Blood tests and other tests may be used for a number of reasons.

1. Blood tests can show if the blood vessels are inflamed. These tests can also measure how the vasculitis is developing, so they may be repeated. A full blood count (FBC) is one of the tests which can be useful. It can help your doctor decide whether you have anaemia (a lack of *haemoglobin*, which is needed to make red blood cells) and whether you have normal levels of white blood cells (which fight infections) and platelets (which clot the blood). Another blood test is the erythrocyte sedimentation rate (ESR), which measures how quickly the cells in the blood settle when they are left to stand in a test tube. This can, indirectly, give an idea of how bad the inflammation is. The C-reactive protein (CRP) test also gives an indication of inflammation. Blood

tests for antineutrophil cytoplasmic *antibodies* (ANCA) are important in the diagnosis of some types of vasculitis, particularly the condition Wegener's granulomatosis.

2. Tests may be carried out to see how the affected body organs are working, such as the kidney (urea and electrolytes and urine tests), or liver (liver function tests). X-rays can test whether the chest is involved. Specific blood tests (such as the CKMB enzyme level) can show whether the heart is involved. The heart can also be assessed by a special ultrasound test (known as an echocardiogram) and an electrical test (the electrocardiogram, or ECG).

3. If your doctor believes that certain organs have been affected but is not sure whether this is due to vasculitis, a biopsy may be carried out. A biopsy means that a small piece of tissue is removed from the organ in question for examination or testing. This is particularly useful to find out if the kidney, the muscles, or the lungs are involved. An ear, nose and throat (ENT) assessment is sometimes carried out for people with Wegener's granulomatosis who have symptoms in these parts of the body.

4. If you are unwell but no specific organs are damaged, then you will sometimes be given an angiogram. This means injecting dye into your arteries so that they show up on an x-ray. This is often done where the abdominal organs such as the kidney and gut are involved. Immunological blood tests can also be very helpful when the diagnosis is uncertain.

5. If you have vasculitis along with other diseases, such as rheumatoid arthritis and systemic lupus erythematosus (SLE), then blood tests may also 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* and antibodies in systemic lupus erythematosus.

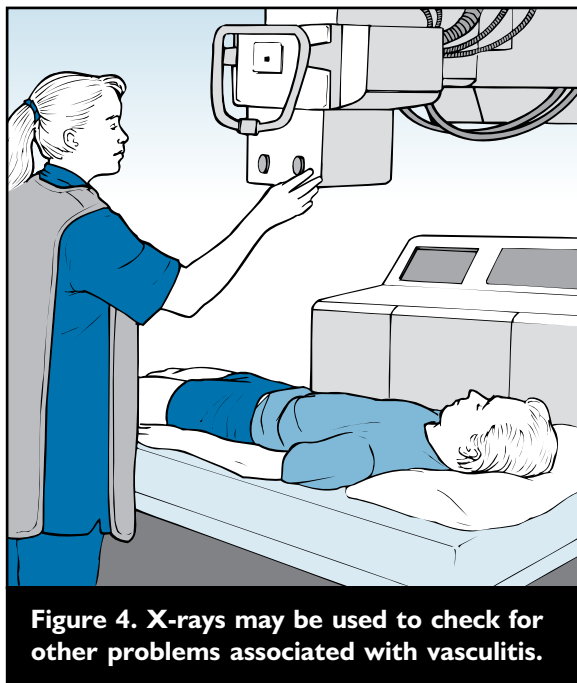


Figure 4. X-rays may be used to check for other problems associated with vasculitis.

For more information on the tests which may be carried out see **arc** booklet 'Blood Tests and X-Rays for Arthritis'.

How can vasculitis be treated?

Treatment has improved a lot in recent years. The type of treatment will depend upon:

- which blood vessels are involved
- how much body tissue is affected
- which body organs are affected.

When an important body organ such as the kidney is involved you will need much more treatment than if you have vasculitis which only affects your skin. The most serious types of vasculitis involve both medium and small arteries.

If you have small-vessel vasculitis, then you may only need a small dose of steroids to control it, if treatment is needed at all. Other drugs such as sulphones may also help. If you have small-vessel vasculitis which only affects the skin, it may be that the only action needed is to treat any underlying infection or to remove the drug or food that triggered the vasculitis.

If you have vasculitis which could affect the kidney (especially if it involves both small and medium-sized blood vessels), then you will probably be given a combination of drugs including:

- an immunosuppressive agent (usually cyclophosphamide), which will suppress the immune system that is attacking the vessels
- corticosteroids, which can be taken as tablets (orally) or given by injection (intravenously).

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

- Kawasaki disease can be treated effectively with injections of globulin (a type of protein).
- Hepatitis-associated polyarteritis nodosa can be treated with anti-viral treatment and thinning or cleaning of the blood (plasma exchange).

If you have vasculitis affecting the large blood vessels, then you will most likely be given corticosteroid tablets. These are very effective for temporal arteritis and Takayasu's arteritis.

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. Because of these risks cyclophosphamide will be discontinued or exchanged for a different immunosuppressive drug, azathioprine, as soon as the vasculitis is controlled. (See **arc** leaflets

'Cyclophosphamide' and 'Azathioprine'.) Possible side effects of corticosteroids include weight gain, indigestion, diabetes, thinning of the skin and thinning of the bones (osteoporosis). If high doses of corticosteroids are given, then you will also be given drugs like bisphosphonates to prevent osteoporosis. (See **arc** leaflet 'Steroid Tablets' and booklet 'Osteoporosis'.)

In some types of vasculitis (e.g. Wegener's granulomatosis) an infection may trigger a relapse. You may therefore be given anti-bacterial drugs such as co-trimoxazole to protect against this. These drugs can also help to protect against the complications of the stronger immunosuppressive drugs.

What can I do to help myself?

If you think you may be developing vasculitis you must go to your doctor as soon as possible. As infections, drugs and foods can sometimes cause the problem, try to think back through the previous few weeks for anything unusual you have taken or eaten. The doctor will probably arrange blood and urine tests.

If you do need treatment then it is very important that you follow the doctor's instructions carefully. You will not usually need to keep to any particular diet, unless the vasculitis has been triggered by a food. If you start on steroids, you should make sure you do not put on too much extra weight (see 'Diet' below). You should also keep a good supply of tablets to make sure the treatment is not interrupted.

The suggestions below may be helpful. However it is very important to recognize that vasculitis can vary enormously from one person to the next and from one type of vasculitis to another. You should therefore ask your doctor or another member of the healthcare team about any new symptoms or anything you are unsure about.

Diet

A healthy, balanced diet is important for everyone, but if you are on steroids it is particularly important because these can increase your appetite and cause weight gain. Try not to eat too much, and cut down on fatty and sugary foods. Instead, eat more fresh fruit and vegetables and starchy foods like bread, potatoes, rice and pasta. Steroids can also make osteoporosis (thin bones) more likely, so it is important to get enough calcium. Foods which are good sources of calcium include tinned sardines (with bones), skimmed milk, yoghurt and certain vegetables such as broccoli. (See **arc** booklets 'Diet & Arthritis' and 'Osteoporosis'.)

Exercise

Vasculitis can cause tiredness. It is important to recognize this and to allow yourself to rest. However, you should also try to keep muscles and joints healthy by exercising.



Figure 5. Try to eat a healthy, balanced diet and avoid putting on extra weight.

Do what you feel you can do, starting, if necessary, with a very small amount of exercise and increasing slowly. Include weight-bearing exercise (such as walking) but you may also find that swimming is an enjoyable way of exercising. Ask your doctor for advice on how much exercise you should expect to be able to do.

Smoking

Avoid smoking. It makes the blood vessels constrict (become narrower inside) and can therefore make vasculitis symptoms worse.

Cold weather

Wearing warm clothes, including warm socks and gloves, helps keep the blood vessels dilated (wider open) by keeping the body warmer. If your fingers feel cold and turn blue, this may be Raynaud's phenomenon. Ask your doctor about this. (More information is given in the **arc** booklet 'Raynaud's Phenomenon'.)

Your doctor or specialist nurse may be able to give you ideas on other ways to help yourself, or recommend a self-help or support group for vasculitis.

What does the future hold?

In the past, some types of vasculitis were very serious diseases, especially if it affected both small and medium-sized arteries. Treatment over the last two decades has completely altered this – 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 (see 'How can vasculitis be treated?').

New research is looking at treatments which will be better than the present ones. Studies with anti-cytokine

drugs (a form of immune treatment) such as infliximab or etanercept may help patients with certain types of vasculitis, including Wegener's granulomatosis and Takayasu's arteritis. Treatment with intravenous proteins (called immunoglobulins or gamma globulins) can also help now.

Researchers are also trying to understand better what causes vasculitis. Comparing what happens in different countries may help to find the answer – for example:

- Takayasu's arteritis is much more common in the East and in Asia than in the UK.
- Temporal arteritis is extremely rare in India but very common in northern Europe.
- Wegener's granulomatosis is more common in northern Europe than in southern Europe.
- Classical polyarteritis nodosa is more common in southern Europe than northern Europe.

arc funds a range of research into vasculitis, including studies on treatments and on the causes and frequency of vasculitis. Recent examples include clinical trials of higher doses of cyclophosphamide, and research into the effects of environmental factors. If differences such as those above can be understood, this will help researchers to develop better treatments.

What are the different types of vasculitis?

Takayasu's arteritis

This affects the main artery from the heart and its large branches, usually in younger women. It is extremely rare in the UK but is more common in Eastern Europe, the Far East and in Africa. The major arteries narrow and this reduces the blood supply to the limbs and other parts of the body. However, the narrowing develops slowly

and the arteries do not usually block completely (see Figure 2). As a result, there is rarely a dangerous loss of blood supply to the arms or legs or any major organs. Steroid treatments are usually effective for this condition.

Temporal arteritis (or giant cell arteritis)

This affects the large arteries which supply the head and neck, especially the temporal artery (over the temple). It is more common in northern Europe, and in people over the age of 50 or 60. As many as 10 people per 100,000 aged over 60 can develop temporal arteritis each year (that is, new cases). It frequently causes headaches. The muscles are often inflamed and stiff in the shoulders and hips (this is polymyalgia rheumatica (PMR) – see **arc** booklet 'Polymyalgia Rheumatica').

IMPORTANT NOTE: Temporal arteritis occasionally involves the blood supply to the eye where it can cause blindness. Fortunately, this risk is very much reduced by steroids and may be further reduced by the addition of aspirin. However, if you have this condition and you have any problems with your eyes



Figure 6. Headaches can be caused by temporal arteritis.

such as blurring or double vision, you should report this to your doctor immediately.

Polyarteritis nodosa (PAN)

This condition is now known as classic polyarteritis nodosa to distinguish it from microscopic polyangiitis (see below). The medium-sized arteries are inflamed, 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. Alternatively, it can involve the whole wall of the artery at a particular point, which causes a blockage. This serious disease is fortunately rare – less than five people per million per year develop it (this is the number of new cases).

Kawasaki disease

Kawasaki disease was first described in Japan in the 1960s. It affects small and medium-sized arteries in young children. It is also sometimes called mucocutaneous lymph node syndrome (because it involves the *mucous membrane*). Children with Kawasaki disease often feel generally unwell – they may have a high temperature, swollen glands in the neck (lymphadenopathy), an inflamed area around the eye (conjunctivitis) and the mouth, and a skin rash which is similar to measles.

Although relatively rare, this condition can be serious if the arteries supplying the heart are inflamed. Up to 60 per cent of patients with Kawasaki disease have this problem (coronary arteritis). Aneurysms can be detected with a special ultrasound technique and angiography. Fortunately, the inflammation often settles with treatment but a few children can die if the aneurysms burst.

Wegener's granulomatosis

This is relatively rare – it affects approximately ten people per million per year and is 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 a more general vasculitis starts. This general vasculitis usually involves the skin, lungs, eyes and kidneys.

Many people with this disease have kidney problems. If this is not recognized early in people with Wegener's granulomatosis, it may lead to kidney failure, which will need to be treated with dialysis. Blood tests can help to show if the kidney is involved and a new test known as ANCA is often used. Some patients have bleeding from the lungs, which causes the coughing up of blood mentioned above.

At one time, Wegener's granulomatosis was a fatal disease. Nowadays, treatment is very effective, especially with a drug called cyclophosphamide. In most patients this can induce long-term remission which can last for many years. Sometimes anti-bacterial drugs such as cotrimoxazole may be given to people with this condition to protect against infections which could trigger a relapse.

Churg–Strauss syndrome

Churg and Strauss were two American pathologists who described this condition. In it, asthma develops in adult life and then inflamed blood vessels are caused by swellings (called granulomas). There will also usually be a high number of *eosinophils* (a particular type of white cell) in the blood.

This is different from Wegener's granulomatosis because of the allergic history (particularly asthma). There is rarely any damage to the ears and nose. Churg–Strauss syndrome more often affects the nerves, which causes weakness, pins-and-needles or numbness.

There is also a higher risk of the heart being involved, which it is important for the medical team to recognize using blood tests and electrical tests (electrocardiogram (ECG) and echocardiogram). If the heart is involved, this usually means there are high numbers of eosinophils in the blood, which can occasionally cause damage (necrosis) to the heart muscle similar to the damage which occurs in a heart attack. Any suggestion that the heart may be involved therefore means that treatment should be given as early as possible.

Microscopic polyangiitis

It is most often kidney specialists who diagnose this – almost all people with this condition have kidney involvement and can develop kidney failure. Patients usually complain that they are ‘always tired’ – this is because of anaemia. Blood tests will show that the kidney is inflamed. The first signs of an inflamed kidney can be spotted by special tests of the urine, which usually show the presence of blood and/or protein.

Because the kidney can also be involved in Churg–Strauss syndrome and Wegener’s granulomatosis, patients must have regular urine tests for blood and protein when all these diseases are considered. Microscopic polyangiitis can also involve the lungs, with bleeding which can cause anaemia and breathlessness.

Henoch–Schönlein purpura

Henoch–Schönlein purpura (HSP) is a rare inflammatory disease of the small blood vessels (capillaries). The exact cause is not fully understood, but research suggests it may be an autoimmune disease. It often follows an acute respiratory infection and may be some kind of allergic reaction to a virus, or in some cases to food or drugs. It mostly affects children aged 2–10 years, though people of any age can be affected. Boys are affected more often than girls, and the older the child (or adult) the higher the risk of it affecting internal organs as well as the skin.

The disease can begin abruptly or may appear gradually over several weeks. Children with HSP all develop a skin rash. The spots are red initially but develop into a bruised purple colour. Each spot lasts for about a week, but they often appear in crops over a period of several days or even weeks.

Children may also have arthritis, especially of the larger joints. Although painful this usually resolves within a few days without any deformity. Some children will have abdominal pain and/or vomiting and can pass bloody stools. Other symptoms include fever, headaches and loss of appetite. Occasionally blood is seen in the urine. This indicates that the kidneys are affected, which is quite common, but serious kidney damage is rare. Occasionally other blood vessels are involved, and serious complications which can occur include bowel perforation, haemorrhage, seizure and stroke, but these are all very rare.

Most children recover fully, although relapses can occur after a disease free interval of several weeks. Relapses are possible for up to a year after the original illness.

There is no specific treatment and in most cases the condition resolves itself. However, anti-inflammatory drugs, and occasionally immunosuppressive drugs, may be given if symptoms persist.

Cryoglobulin-associated vasculitis

In this disease, small-vessel vasculitis is associated with cryoglobulins – these are proteins in the blood which stick together in the cold. This is an important disease to recognize because too many cryoglobulins can cause ‘sludging’, which can reduce the flow of blood or even block the blood vessels causing damage to the organs or body tissues. Cryoglobulins can be removed from the blood by a mechanism called plasmapheresis (plasma exchange).

Conclusion

arc is actively involved in supporting research into the cause and treatment of vasculitis in many units in the UK (see ‘What does the future hold?’). There have been important improvements in treatment in recent years and it is to be hoped that further advances will follow as more is discovered about the disease.

Glossary

Antibodies – blood proteins which are formed in the body to respond to germs, viruses or any other substances which the body sees as foreign or dangerous. The job of antibodies is to attack these foreign substances and make them harmless.

Complement – an enzyme ‘system’ in the blood. An enzyme is a substance which speeds up a biological reaction (acts as a catalyst). Complement is a ‘cascading enzyme system’. This means that a number of different enzymes are created or triggered one after another. This then allows a number of reactions to take place. The complement system 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. The blood can be tested to find how much of each of the major elements of complement is present.

Dialysis – a method of separating particles in a liquid using a semi-permeable membrane. In kidney dialysis the patient’s blood is circulated through a special machine which uses this method to remove waste materials or poisons from the blood.

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

Haemoglobin – is found in red blood cells and contains the pigment which gives blood its colour. Because it can combine with, and then release, oxygen, it allows the blood to carry oxygen around the body.

Mucous membrane – the type of membrane which lines the areas of the body such as the mouth, nasal passages, stomach and gut, vagina, and passages to the lungs.

Useful addresses

The Arthritis Research Campaign (arc)

PO Box 177
Chesterfield
Derbyshire S41 7TQ
Phone: 0870 850 5000
www.arc.org.uk

As well as funding research, we produce a range of free information booklets and leaflets. Please contact the address above for a list of titles.

Arthritis Care

18 Stephenson Way
London NW1 2HD
Phone: 020 7380 6500
Helpline (freephone): 0808 800 4050
www.arthritiscare.org.uk

Offers self-help support, a helpline service, and a range of leaflets on arthritis.

National Kidney Federation

6 Stanley Street
Worksop
Notts S81 7HX
Helpline: 0845 601 0209
www.kidney.org.uk

Provides advice and information to kidney patients, carers and medical professionals.

Stuart Strange Vasculitis Trust

12 Acton Road
Mackworth
Derby DE22 4JF
Phone: 01332 521595
www.vasculitis-uk.org

Offers information and support to people with vasculitic disease and their families.

Vasculitis Foundation

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

Produces a newsletter which is available to people outside the USA (there is a subscription fee).

Arthritis Research Campaign



The Arthritis Research Campaign (**arc**) is the only major UK charity funding research in universities, hospitals and medical schools to investigate the cause and cure of arthritis and other rheumatic diseases. We also produce a comprehensive range of over 90 free information booklets and leaflets covering different types of arthritis and offering practical advice to help in everyday life.

arc receives no government or NHS grants and relies entirely on its own fundraising efforts and the generosity of the public to support its research and education programmes.

Arthritis Today is the quarterly magazine of **arc**. This will keep you informed of the latest treatments and self-help techniques, with articles on research, human interest stories and fundraising news. If you would like to find out how you can receive this magazine regularly, please write to: Arthritis Research Campaign, Ref AT, PO Box 177, Chesterfield S41 7TQ.



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