Once the drug doses have been reduced to maintenance levels, usually about 3 months after starting treatment, careful monitoring is required to check that side effects of the drugs do not develop, but also to see if vasculitis comes back (a relapse). An early relapse may be picked up on routine blood tests, but it may also cause some symptoms - usually those that occurred during the early stages of the disease, before treatment was started. If someone suspects a relapse, they should get in touch with their doctor urgently.

The ‘traditional’ drugs used in vasculitis are steroids (prednisolone), cyclophosphamide, and many people are switched from cyclophosphamide to a milder drug called azathioprine after 3-6 months. In some people mycophenolate mofetil is used instead, particularly when azathioprine leads to side effects. Rituximab is a new type of drug which is given in a course of injections which works on a specific group of white cells. This is often used when cyclophosphamide cannot be used and if the disease comes back despite treatment.

In people who have very active vasculitis with kidney damage, there may be a benefit to a treatment called ‘plasma exchange’, or ‘plasmapheresis’. This removes plasma from the blood, which may contain ANCA and other harmful substances. Plasma exchange may be used for a course lasting a week or two.

In some cases the kidneys are so badly damaged before doctors see someone that they will not work again. Treatment with steroids and cyclophosphamide is therefore aimed at treating vasculitis in other parts of the body.

What can I do?
Unfortunately it seems there is nothing you can do in your lifestyle or diet that will stop you getting vasculitis. Similarly, there is little you can do that would prevent you getting a relapse. However, what you do still makes an enormous difference to your health:

1. Remember to take your medication at the times prescribed. Doctors often change the doses of tablets, so make sure you know exactly what has been advised. If in doubt about the dosage of tablets, phone up and check.
2. Report possible complications of treatment such as symptoms of infection at an early stage.
3. Take regular exercise. People with vasculitis lose a lot of muscle strength during illness. Ask for advice about sensible regular exercise.
4. Eat well. You will probably lose some body weight with vasculitis in its early stages. Even if you have a weight problem, this weight loss is usually a bad thing, because it is your muscle and not your fat that is lost! Please take advice from the renal dietitians about regulating your diet properly.

The National Kidney Federation cannot accept responsibility for information provided. The above is for guidance only. Patients are advised to seek further information from their own doctor.

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What is vasculitis?
Vasculitis is a medical term for inflammation of blood vessels. It can be caused by different diseases, however, their treatment is very similar.

The names of some of the various types of vasculitis depends on the size of the blood vessel that is inflamed. Kidneys are often affected by small vessel vasculitis and are called: microscopic polyarteritis (poly-angiitis) and granulomatosis polyangiitis (previously known as Wegener’s granulomatosis).

What causes vasculitis?
The cause is immunological. That is, the white blood cells and antibodies (natural defences) you have to fight off infection are damaging your own body by mistake. The body’s defences against infection are very powerful and many diseases are due to their overactivity such as asthma, rheumatoid arthritis and diabetes in young people.

Vasculitis is different from these other diseases because blood vessels in particular are damaged. Doctors do not understand fully why this happens and the condition is quite rare. Vasculitis with kidney involvement affects 20-30 people per million population per year.

The trigger that starts vasculitis varies from person to person. Sometimes it is a ‘flu-like infection’, sometimes it appears to be an operation or a bacterial infection. The condition is commoner in the spring and autumn than in the summer and is commoner in older people.

What are the Symptoms?
These are variable and almost every case is different from each other. Often the only symptom is a feeling of tiredness and general ill health. Blood vessels in all parts of the body can be affected; here are some of the commoner symptoms:-

Kidneys: the specialised small blood vessels in the kidney which filter blood to make urine seem especially sensitive to vasculitis. The damage to the kidney can cause blood to appear in the urine as well as kidney failure in severe cases. This is a serious condition and when it is suspected doctors would normally recommend a kidney biopsy test to confirm the diagnosis and to help plan treatment.

Skin: small blood vessels in the skin can become damaged by vasculitis, causing them to burst, causing a rash with small red blotches. This is most commonly seen on the lower legs.

Joints: inflammation around the joints can cause pain and stiffness.

Nose: the inside of the nose is often affected by vasculitis, so there can be nosebleeds. These may even occur for some time before the kidneys are affected.

Lungs: bleeding can occur inside the lungs in vasculitis. If you cough up blood, it is important that you are seen and treated urgently.

Other parts of the Body: we see almost every part of the body affected in some cases of vasculitis. It can rarely cause stroke, heart attack or damage to the bowel. Fortunately, once treatment has been started, it is unusual for such problems to start.

How is vasculitis diagnosed?
Vasculitis should be suspected in anyone with a severe, recent onset kidney disease where a urine test shows traces of blood and protein in the urine. If someone has a typical rash on the skin it may be easy to diagnose, but in many cases the diagnosis is delayed because the symptoms of tiredness, and perhaps shortness of breath, may occur in several different conditions which are commoner than vasculitis.

If vasculitis is suspected, blood tests will be performed. The most important is called ANCA (anti-neutrophil cytoplasmic antibodies). ANCA are antibodies against white blood cells and under certain conditions they stimulate white blood cells to damage the kidneys. The ANCA test is very useful in diagnosing vasculitis and in many cases the level of ANCA can be used to monitor someone’s progress. ANCA itself usually consists of another specific antibody with the abbreviations MPO or PR3. If you have vasculitis and one of these is positive, it will be used to monitor disease activity so you might like to ask what the level is at clinic visits. However, some people have vasculitis with no ANCA in the blood.

It is often necessary to look at a piece of kidney under the microscope to confirm whether vasculitis is present. This is removed with a needle, and the test is called a kidney biopsy.

What is the Treatment?
Vasculitis often responds very well to treatment. A combination of steroids (prednisolone) and cyclophosphamide is usually used. These are both powerful drugs that reduce the activity of the immune system, reducing the activity of vasculitis. High doses of these drugs are normally given for the first 3-6 months after which doses are reduced to maintenance levels. Some treatment is needed for at least 2 years and in many cases for the rest of your life.

These drugs can cause serious side effects and very careful monitoring is required for the sake of safety. The main side effect is infection. There is a risk of severe urine infection or pneumonia during the first few months of treatment and any fevers or possible infection should be reported urgently to your doctor.