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# Vasculitis Factsheet

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This is a leaflet for people who want general information about vasculitis. Vasculitis is a medical term meaning inflammation of blood vessels. It can be primary (occurring on its own) or secondary (occurring as part of another condition). Some people with vasculitis test positive for antibodies to constituents of certain white blood cells (anti-neutrophil cytoplasmic antibodies or ANCA) and are said to have ANCA vasculitis.

## Who gets vasculitis?

Vasculitis is an uncommon illness. About 20 in every 100,000 people get ANCA vasculitis every year in the UK. Vasculitis can affect all age groups. Some are mostly diseases of childhood (e.g. Kawasaki), whilst others persist throughout adult life (ANCA systemic vasculitis). Some principally affect the elderly (e.g. giant cell arteritis).

## What causes vasculitis?

In most cases the cause is not known. Many experts think that the illness is the result of infection in people who were born with a certain genetic predisposition.

Vasculitis can also be caused by illnesses which trigger inflammation in the body such as rheumatoid arthritis and inflammatory diseases of the bowel.

Medicines associated with vasculitis include certain types of antibiotics (e.g. quinolones, sulphonamides, beta-lactams), anti-inflammatories, the contraceptive pill, some types of fluid tablets (thiazides) and flu vaccines.

Rare types of cancer (paraproteinaemia, lymphoproliferative disorder) can occasionally cause vasculitis.

## Are there different types of vasculitis?

There are many different types of vasculitis and various ways of classifying them. Vasculitis occurring in one part of the body is known as localised vasculitis. Vasculitis in several parts of the body is known as systemic vasculitis. There are nearly twenty types of primary systemic vasculitis.

The commonest types of vasculitis are:

- ANCA\* associated vasculitis including microscopic polyangiitis, Granulomatosis with polyangiitis, Churg Strauss
- Giant cell arteritis
- Polyarteritis nodosa
- Henoch Schonlein purpura
- Kawasaki

One way of classifying vasculitis is by the size of blood vessel they affect, e.g. small, medium or large. Other ways of identifying different types include the parts of the body affected, the features seen on tissue biopsy and the pattern of blood test findings.

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### What are the symptoms of vasculitis?

To start with, you may feel generally unwell and get fever, night sweats, weight loss and tiredness. After that, the symptoms will depend on the area involved. You may notice a rash if the inflammation affects the blood vessels of the skin. Involvement of the nose may cause stuffiness, nose bleeds and sinus pain and involvement of the lungs can result in breathlessness and coughing up blood. Deafness and ear pain can occur.

Depending on the type of vasculitis, you may get joint or muscle pains, headaches or tingling of the arms and legs. Vasculitis in less obvious areas of the body such as the kidneys may not cause any symptoms in the early stages. It may only show up if the urine is tested as part of a health check and found to contain blood or protein. Long-term vasculitis of the kidneys can lead to kidney failure.

### How is vasculitis diagnosed?

Vasculitis may be difficult to diagnose but should always be borne in mind in people who have unexplained symptoms. Blood and urine tests are usually helpful. A chest x-ray should be done if lung disease is a possibility. Other tests will depend on the type of vasculitis suspected. For example, a skin biopsy may be required to diagnose a rash, and kidney biopsy may be necessary if there is an unexplained finding of blood or protein in the urine.

### What is the treatment for vasculitis?

Steroids are usually prescribed. They are normally continued for many months after disease is brought under control. You will most likely also be offered a medicine which reduces the activity of the immune system (immunosuppressant), such as cyclophosphamide, azathioprine or methotrexate. You will most likely need to take this medicine for a long time (months or years).

### Does vasculitis get better?

With the help of modern medicine, you stand a very good chance of having your symptoms brought under control. If your condition is treated early enough the risk of getting long-term damage is low. Occasionally if treatment is delayed, complications such as kidney damage can occur and can shorten life span. Other illnesses associated with vasculitis include heart disease, stroke and rarely cancer.

### Further reading

Luca N et al, Vasculitis and Thrombophlebitis, May 2012

Heeringa P et al In vivo approaches to investigate ANCA-associated vasculitis: lessons and limitations, 2011.