

## Vasculitis disorders

Vasculitis disorders result from inflammation of blood vessels, are relatively rare and can affect people of all ages. Granulomatosis with polyangiitis (previously known as Wegener's granulomatosis) is the most common form of vasculitis and affects around 5 in a million people.

Whilst some vasculitis disorders improve on their own, most require treatment. Vasculitis disorders have many clinical presentations, sometimes affecting only the skin but also internal organs. Due to the wide variation in symptoms, the length of treatment varies, with some people using medications for extended periods of time.

### What is vasculitis?

Vasculitis disorders result from inflammation of blood vessels, including arteries, arterioles, veins, venules and capillaries. The inflammation causes a narrowing of blood vessels, which can result in blood flow obstruction (ischaemia). This may lead to tissue damage (necrosis) and blood clots (thrombosis). The different types of vasculitis are classified according to the size and location of the blood vessels that are affected.

### What causes vasculitis?

There are three main underlying causes of vasculitis disorders:

- Autoimmunity
- Allergy or hypersensitivity to medications, toxins or other inhaled environmental irritants - elimination of the medication, toxin or irritant usually results in cessation of symptoms.
- Viral or parasite Infections

Allergy, hypersensitivity and infections should be ruled out before autoimmunity is considered as the cause of a vasculitis disorder.

### Autoimmune related vasculitis

The main role of the immune system is to fight foreign invaders such as bacteria, moulds and viruses. In autoimmune diseases the immune system produces antibodies that attack the body's own healthy tissue. Antibodies produced by the immune system in vasculitis disorders cause inflammation in blood vessels that can lead to serious problems. Complications depend on which blood vessels, organs and other systems are affected.

Vasculitis disorders may also occur in people with other autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematosus (SLE) and dermatomyositis.

### Diagnosis

The diagnosis of vasculitis disorders is usually suspected on the basis of clinical symptoms and signs, confirmed by laboratory tests. An accurate diagnosis is required to provide appropriate treatment.

Due to the wide range of signs, symptoms and body systems involved, an extensive history and physical examination is required before a diagnosis of the type of vasculitis disorder can be made. A range of blood tests to find evidence of autoimmunity are usually performed. In some cases an x-ray of the blood vessels or a biopsy of the affected organ/s may be recommended.

### Treatment

It is important to find out as much as possible about your condition by asking questions of your clinical immunology/allergy specialist or other treating doctor.

When vasculitis is due to an autoimmune disorder, immunosuppressive drugs are usually used.

Plasmapheresis (a procedure that filters the offending autoantibodies out of the blood plasma and returns the filtered blood back to the patient) may be used in serious cases that do not respond to other treatment.

### Signs and symptoms

General signs and symptoms of vasculitis disorders include:

- Fever
- Loss of appetite and weight loss
- Fatigue, weakness and lethargy
- General aches and pains

Specific signs and symptoms of vasculitis disorders include:

- **Skin** - purple or red spots or bumps, clusters of small dots, splotches, bruises, urticaria (hives), itch.
- **Joints** - aching joints, arthritis in one or more joints.
- **Lungs** - shortness of breath, coughing up blood, signs that suggest pneumonia.
- **Gastrointestinal tract** - mouth ulcers (sores), stomach pain. In severe cases blood flow to the intestines can be blocked.
- **Sinuses, nose, throat, ears** - chronic (ongoing) sinus or middle ear infections, ulcers in the nose, hearing loss.
- **Eyes** - red, itchy and burning eyes, increased sensitivity to light, blurred vision. Severe cases can result in blindness.
- **Brain** - headaches, changes in mental function, stroke-like symptoms such as muscle weakness and paralysis.
- **Nerves** - numbness, tingling, and weakness in various body parts of your body, loss of feeling or strength in hands and feet, shooting pains in arms and legs.

### Examples of vasculitis disorders (small blood vessels)

- **Granulomatosis with polyangiitis** (the most common form of vasculitis) - affects sinuses, lungs, kidneys, skin (previously known as Wegener's granulomatosis)
- **Eosinophilic granulomatosis with polyangiitis** - affects lungs, skin, nerves (previously known as Churg-Strauss syndrome)
- **Cryoglobulinaemia** - affects skin, kidneys, nerves
- **Goodpasture's syndrome** - affects lungs, kidneys
- **Henoch-Schonlein purpura** - affects skin, joints, kidneys, gut
- **Microscopic polyangitis** - affects skin, kidneys, nerves

### Examples of vasculitis disorders (medium blood vessels)

- **Behcet's disease** - affects mucous membranes, skin, eyes
- **Central nervous system vasculitis** - affects brain
- **Kawasaki syndrome** - affects skin, mucous membranes, lymph nodes, blood vessels
- **Polyarteritis nodosa** - affects arteries, kidneys, gut, nerves, skin

**Examples of vasculitis disorders (large blood vessels)**

- **Giant cell (temporal) arteritis** - affects arteries of the head and neck
- **Takayasu arteritis** - affects arteries of the head and neck
- **Polymyalgia rheumatica**

**Further information**

There are organisations which provide support and information to people with autoimmune disorders. These organisations are listed on the ASCIA website [www.allergy.org.au/patients/patient-support-organisations](http://www.allergy.org.au/patients/patient-support-organisations)

The following websites may also be useful:

[www.vasculitisfoundation.org](http://www.vasculitisfoundation.org)

[www.vasculitis.org.uk/](http://www.vasculitis.org.uk/)

[www.nhlbi.nih.gov/health/health-topics/topics/vas](http://www.nhlbi.nih.gov/health/health-topics/topics/vas)

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