



# Vasculitis - The Facts

***This factsheet is intended as a simple introduction to vasculitis for those who have just been diagnosed with vasculitis, members of their family, friends, work colleagues and for others who may want to know about the disease.***

## **What is Vasculitis**

Vasculitis is a rare inflammatory disease which affects about 2-3000 new people each year in the UK. Vasculitis means inflammation of the blood vessels. Any vessels in any part of the body can be affected.

There are several different types of vasculitis. In the first type, the acute form, it can be caused by infections, reaction to drugs or exposure to chemicals. Often the problem is localised, such as a rash. In these cases the disease usually needs no treatment. Other types of vasculitis can be secondary to (or as a consequence of) other illnesses such as rheumatoid arthritis or some types of cancer.

The third group is known as Primary Systemic Vasculitis (PSV). Primary means that it is a disease in its own right, not secondary to any other illness. Systemic means that it can involve any part of the body.

Primary Systemic Vasculitis is an auto-immune disease with no known cause, although genetic makeup may make some people more susceptible.

In PSV, the body's own white blood cells (which normally defend us by attacking and removing "foreign" cells in the blood stream) start to attack the cells lining the blood vessels. This can cause partial blockage of larger vessels but complete blockage of medium and small arteries, veins and capillaries. This in turn leads to death of the tissues supplied by those vessels.

There are various types of PSV. Some are mild and require no treatment, but others are very severe and cause serious illness, even death, if not recognised and diagnosed promptly and treated appropriately and effectively.

## **Symptoms and Treatment**

Primary Systemic Vasculitis can affect various organs, and the symptoms of vasculitis are easily confused with other illnesses, so the disease can go unrecognised for a long time.

The organs that can be affected may be the upper respiratory system (nasal passages and sinuses) and the lower respiratory system (trachea and lungs), ears, eyes, skin, kidneys, gastro-intestinal system, nervous system, and others. So different types of vasculitis can cause loss of sight, deafness, loss of digits or limbs, skin scarring and facial disfigurement, kidney failure, damage to lungs, trachea, nasal passages and sinuses, gastro-intestinal disorders, damage to the brain and nervous system and occasionally to the heart.

There is no cure for PSV, but it can usually be controlled by use of steroids, chemotherapy and immune suppressing drugs. Long term drug therapy is often required. If all goes well some patients go into "full remission" - ie they no longer need drugs. But relapse is common.

People suffering from vasculitis often experience muscle weakness and chronic fatigue. Some experience chronic pain due to nerve damage or severe migraines and headaches due to damaged blood vessels in the head.

Others require dialysis or kidney transplants. Many have breathing problems and others are left with permanent physical disabilities.

## **The most "common" types of these rare vasculitis diseases are:**

- Granulomatosis with Polyangiitis (GPA) (previously known as Wegener's Granulomatosis)
- Eosinophilic Granulomatosis with Polyangiitis (EGPA) (previously known as Churg Strauss Syndrome)
- Giant Cell Arteritis/Temporal Arteritis (GCA)
- Henoch Schönlein Purpura (HSP)
- Microscopic Polyangiitis (MPA)
- Polyarteritis Nodosa (PAN)

## **Rare types of vasculitis include:**

- Behçet's Disease (BD)
- Central Nervous System Vasculitis or Cerebral Vasculitis (CNS)
- Cryoglobulinemia
- Kawasaki's Disease (KD)
- Takayasu's Arteritis (TA)
- Urticarial Vasculitis (HUVs)

The key to successful treatment is early recognition and early correct diagnosis, followed by prompt, appropriate and effective treatment. This results in better subsequent quality of life and longer life expectancy

## **Caring for a vasculitis patient**

On the Vasculitis UK website there is a section devoted to caring for a vasculitis patient. This is useful information for carers, families and friends. See: [www.vasculitis.org.uk/living-with-vasculitis/caring](http://www.vasculitis.org.uk/living-with-vasculitis/caring)

For much more information about the individual vasculitis diseases, about vasculitis in general and how to live and cope with vasculitis visit our website:

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

## **CONTACT DETAILS**

John Mills, West Bank House, Winster. DE4 1DQ  
e-mail: [john.mills@vasculitis.org.uk](mailto:john.mills@vasculitis.org.uk)  
Phone: 0300 365 0075