



# Route Map for Vasculitis

## Introduction

The term "Vasculitis" covers a range of varied and quite dissimilar diseases. Vasculitis means literally "inflammation of the blood vessels" and may include everything, from a mild allergy type reaction (caused perhaps by exposure to a drug, a chemical or something in nature like an insect bite), to a serious systemic multi-organ auto-immune disease that can be life threatening. For that reason, all types of vasculitis should be regarded as a very serious and potentially fatal disease until proven otherwise.

Although systemic vasculitis is incurable it can be controlled by modern medication. Provided that the disease is recognised and diagnosed correctly at an early stage and is then treated appropriately and effectively, most patients make a good recovery and with adequate ongoing care, continue to have a good quality of life and normal life expectancy.

Systemic vasculitis is an uncommon, highly complex, very variable and often unpredictable chronic disease. It is often difficult to diagnose and disease activity may be very difficult to assess and predict. Clinical signs and symptoms are common to many other diseases. The results of laboratory and other investigations are often not specific to vasculitis or in some cases, the results can be misleading. So differential diagnosis is difficult.

It is often the case that a correct diagnosis will only be reached by a clinician who has considerable experience of vasculitis. Effective diagnosis necessitates the assessment of **all** the clinical signs and symptoms alongside the results from investigations. A multi-disciplinary approach is essential. For this reason, cases should not be treated by clinicians who do not have adequate knowledge and experience of vasculitis, its diagnosis and management unless they also have support and guidance from a doctor in an acknowledged centre of expertise who is familiar with dealing effectively with vasculitis in its diverse forms.

Doctors dealing with cases of suspected vasculitis should be aware that for ANCA associated vasculitis, the one and five year survival rates are statistically worse than those for both breast and prostate cancer.<sup>1, 2, 3</sup> . Much of the mortality occurs in the early stages of the disease, due to late recognition and diagnosis. Systemic vasculitis may also cause irreversible damage to single or multiple organs unless it is recognised early, diagnosed promptly and treated properly and effectively.

For the above reasons, it is essential that the early signs of vasculitis should be recognised and investigated thoroughly. Diagnosis should be prompt and treatment should be effective and carefully monitored. In most cases, treatment involves an initial induction phase followed by a prolonged period of maintenance.

Under-treatment may result in further or continuing organ damage or early relapse. Over treatment may result in excessive immune suppression and consequent risk of serious opportunistic infections. Medication used in treating systemic vasculitis often has serious side effects so there is the need for constant regular, considered monitoring. It is a matter of carefully balancing the need for effective control of the disease against the adverse side effects of the treatment. For these reasons clinicians who have little experience of managing different types of vasculitis cases would be well advised to seek advice from colleagues who do.

Systemic vasculitis is a chronic disease, which is controllable but at present is not curable. It is frequently subject to relapse. Patients suffering from most types of vasculitis, even when apparently in complete remission, cannot safely be completely discharged from care and should continue to be subject to periodic monitoring.

Clinicians and patients should also be aware of the latest guidelines on the diagnosis and treatment of vasculitis. Links to these are shown in this Route Map and can readily be found on the Vasculitis UK website [www.vasculitis.org.uk](http://www.vasculitis.org.uk) under the "Professionals" heading.

This Vasculitis Route Map is only an introductory guide to the many different types of vasculitis. However there are links to other sources that offer more information. The full content of this Route Map is also available in the pages of the Vasculitis UK website [www.vasculitis.org.uk](http://www.vasculitis.org.uk) where it is further expanded and is constantly updated.

### References:

<sup>1</sup> Flossman O, Berden A, de Groot K et al. "Long Term Patient Survival in ANCA-associated vasculitis". Ann Rheum Disease. 70.488-494, 2011

<sup>2</sup> Cancer Research UK, Breast cancer-survival statistics, weblink:  
<http://info.cancerresearchuk.org/cancerstats/types/breast/survival/>

<sup>3</sup> Cancer Research UK, Prostate cancer-survival statistics, weblink:  
<http://info.cancerresearchuk.org/cancerstats/types/prostate/?script=true>

John Mills (Chairman, Vasculitis UK) November 2012

### Comments on the Route Map for Vasculitis from Dr David Jayne and Professor Lorraine Harper:



"I have been most impressed by the scope and consistent quality of the vasculitis route map. The lack of availability of reliable information for vasculitis patients and their carers continues to be another hurdle in the journey they have to make, and the route map will be widely welcomed. For a potentially severe disease, such as vasculitis, an understanding of what is known, and not known, and of what lies ahead enables patients to better participate in medical decisions about their future. Promoting a partnership between patients and health care professionals will drive up standards of care."

**Dr David Jayne**

Consultant - Vasculitis & Lupus Clinic  
Cambridge University Hospitals NHS Foundation Trust



"The diseases we call vasculitis are rare but have a big impact on the people they affect. We hope the Route Map will become a reference source for people affected by any kind of vasculitis and the GPs who care for them."

**Prof Lorraine Harper**

Professor of Nephrology, Birmingham University Hospital and the medical advisor to Vasculitis UK



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## **Amendments and Additions since first publication**

**See Contents page for changes to page Nos.**

### **Additions up to May 2013**

Anti-GBM (Goodpastures Disease)  
Introduction  
Urticarial Vasculitis  
Vasculitis and the Ear  
Vasculitis and the Eye

### **Revisions at October 2013**

Aims of the Trust (Trustee details)  
Articles in the Vasculitis UK Newsletter/Journal  
Disability Living Allowance  
Personal Independence Payments  
Vasculitis Support Groups  
Web links

### **Revisions at July 2014**

Low Mood and Depression  
Pain Management  
Understanding Feelings and Reactions to Illness  
Behcet's Syndrome  
Takayasu Arteritis  
Articles in the Vasculitis UK Newsletter/Journal

The Route Map for Vasculitis has been produced by Vasculitis UK in conjunction with the Renal Immunobiology Group at the University of Birmingham. This work is part of the Route Maps for Rare Conditions project, facilitated by Genetic Alliance UK and funded by the Department of Health in England.



**With special acknowledgements to:**

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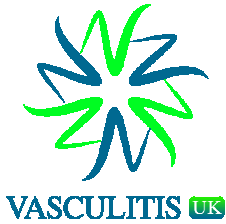
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**The Trustees of Vasculitis UK (2011/12)**  
Paul Bingham, Andy Bone, Pat Fearnside, Mark Griffiths, Holly Hampshire,  
John Mills, Susan Mills, Chris Stait, and Martin Thomas

**Jessica Burke**  
Genetic Alliance UK

Since the Route Map was first published further articles have been added and we would like to thank the following for their contributions:

**Dr David Jayne (Fertility and Vasculitis), Dr Catherine Guly (Vasculitis and the Eye),  
Dr Marcos Martinez Del Pero (Vasculitis and the Ear)**



## About Vasculitis UK

The Trust was officially re-named "Vasculitis UK" in Spring 2011, having originally been known as the "Stuart Strange Trust" and later "Vasculitis UK (Stuart Strange Vasculitis Trust)".



**Our roots:** The Trust was originally set up by the family and friends of Stuart Strange. Stuart died prematurely of Wegener's Granulomatosis in 1992, aged 29.

Originally the Trust concentrated on fundraising for and raising awareness of Wegener's Granulomatosis but soon realised that there were patients suffering from other forms of vasculitis, and the range was widened to include all the vasculitic diseases. Stuart's family wanted to do everything they could to help patients being diagnosed with vasculitis. At the time of Stuart's illness and subsequent death there had been no help for the family. His wife and sister wanted to ensure that that if they could only give one other person some degree of comfort then it was going to be a worthwhile cause.

Charity status was obtained during 1993. Stuart's mother, Lillian, is the Life President of the Trust.

**The present:** Vasculitis UK is the No. 1 vasculitis charity in the UK. Membership of the Trust has grown considerably over the years. In addition the number of independent regional support groups has also increased. These independent groups help the Trust to achieve its aims by holding group meetings - some informal and some with eminent vasculitis speakers in attendance.

The Trust offers support to vasculitis sufferers, their families and friends both at home and abroad. In addition the Trust aims to increase awareness of vasculitic diseases among both the general public and health professionals. Vasculitis UK also supports research into the causes of and treatments for vasculitis.

The Trust is financed solely by **voluntary** donations received from members, friends and family, other supporters, fundraising events and bequests made in memory of loved ones. There are no joining or annual fees. Vasculitis UK is administered by nine unpaid Trustees. There are no offices and no paid staff.

How does the Trust spend the donation received? On average:

- Approximately 15% is spent on printing, postage and incidental expenses,
- Approximately 5% is spent on information booklets and leaflets.
- The remainder goes to fund research into the causes of and treatments for vasculitis. The Trust has donated over £125,000 during the past five years to fund research projects at Birmingham University Hospital and Addenbrooke's Hospital here in the UK.

**Registered Charity No. 1019983**



## Aims of the Trust

Vasculitis UK is an independent organisation funded only by voluntary contributions from members and supporters.

The aims of the Trust are:

- To offer support and advice for those with vasculitis and their families
- To support and promote research into the causes and treatments of vasculitis
- To increase awareness of vasculitic diseases among both the general public and health professionals
- To support the development of local vasculitis support groups

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

**Vasculitis UK - formerly known as Stuart Strange Vasculitis Trust**

**Registered Charity No. 1019983**



## About Genetic Alliance UK

Genetic Alliance UK is the national charity of patient organisations with a membership of over 140 charities supporting all those affected by genetic disorders. The aim of Genetic Alliance UK is to improve the lives of people affected by genetic conditions by ensuring that high quality services and information are available to all who need them.

Genetic Alliance UK seeks to improve the lives of people affected by genetic conditions by ensuring that high quality services and information are available to all who need them. The aims of Genetic Alliance UK include:

- **Supporting:**  
By raising awareness of genetic diseases and improve the quality of services and information available to patients and families.
- **Campaigning:**  
Actively campaign on issues of policy and practice to influence governments, policy makers, industry and care providers such as the National Health Service.
- **Uniting:**  
Providing a united voice for all those affected by genetic conditions, enabling us to work together towards a common goal of making life better for patients and families at risk.

## About the Route Maps for Rare Conditions Project

The Route Maps for Rare Conditions project was developed as a practical and cost-effective framework for improving information, access and coordination of health and social care services for individuals and families with a wide range of rare genetic conditions. Nine rare disease groups have produced a Route Map for their own disease.



Each Route Map provides a comprehensive resource for use by patients, families and health and social care professionals and these have the potential to play a key role in personalised care planning for people with these conditions.

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

**Registered Charity Nos: England & Wales 1114195, Scotland SC039299**



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## **A wise man knows one thing - the limits of his knowledge**

**John Maynard Keynes**

### **Who is this Route Map for?**

- **Patients and families**

The pages of this Route Map contain details of the various vasculitic diseases, symptoms, treatments, prognosis etc. There are also sections covering government benefits, prescription charges, sources of information and much more for the benefit of the vasculitis patient and their families and also for medical practitioners. The aim of the Route Map is to offer a complete guide to vasculitis in one document.

- **Medical Practitioners**

The Route Map includes information for the benefit of physicians wishing to learn more about the vasculitides. We have included a list of web links to freely available review articles covering the vasculitides included in this Route Map. They provide a basic introduction to the clinical features and treatments at the time of writing this document. They are not comprehensive and anyone needing to learn more about treating vasculitis is advised to review the most up to date evidence available and/or discuss with a consultant experienced in managing the appropriate disease.

There are a number of hospitals with "multi-disciplinary" clinics for the treatment of vasculitis. A list of these hospitals is available by contacting John Mills (Chair, Vasculitis UK) see contacts page for details.

- **Other health, social care and benefit agency professionals**

Included in this document are details of other problems encountered by the patient with vasculitis. The aim is to highlight the difficulties encountered by patients and their families, such as accessing information and assistance with diseases which requires continuous monitoring, drug therapy, and frequent relapse.

## Vasculitis Basics

### What is Vasculitis?

Vasculitis is inflammation of the blood vessels. The blood vessels involved can be in any organs and may be different in different types of vasculitis. The skin, joints, kidney and lungs are often involved in some of the more common types of vasculitis. The overall prognosis varies depending on the organ involved and the severity of the disease. Treatment varies depending on the actual vasculitic disease present, the severity of the disease and on the organs involved. For more information see the individual vasculitis disease sections.

There are different types of blood vessels in the body and each different type of vasculitis will usually affect specific kinds of blood vessel. It may be helpful to have an understanding of the different types of blood vessel to help understand the different types of vasculitis.

The blood vessels that carry blood from the heart to the body's organs are called arteries. The main artery coming out of the heart is called the aorta and from this smaller arteries branch off (like roads coming off a motorway) to go to specific organs (eg the renal arteries go to the kidneys, the carotid arteries go to the head and the coronary arteries supply the heart muscle). Inside the organ the arteries divide up into smaller and smaller arteries called arterioles and finally into the very tiny blood vessels called capillaries (imagine the tiniest veins in a leaf from a tree) where oxygen and nutrients come out of the blood to supply the organ. After this the capillaries join up to form veins to go back to the heart and then on to the lungs to be resupplied with oxygen before going back to the heart and around the body again.

Different types of vasculitis will usually affect mainly one size of blood vessel. Takayasu's arteritis and giant cell or temporal arteritis are called large vessel vasculitis because they mainly affect the aorta and the biggest arteries as they branch off. Polyarteritis nodosa and Kawasaki's disease are called medium vessel vasculitis because they affect the middle sized arteries as they go into the organs. Most of the other types of vasculitis, such as Wegener's granulomatosis, microscopic polyangiitis etc, are called small vessel vasculitis because they cause inflammation in the smallest blood vessels, the capillaries, as they supply the insides of the organs. Vasculitis is an auto-immune disease where the body's immune system attacks blood vessels instead of defending them against infection.

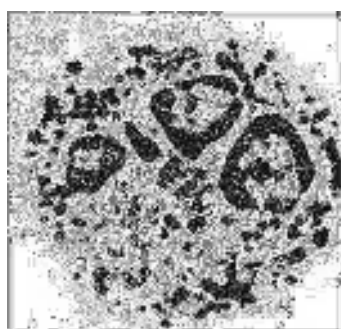
### The Immune System and Autoimmune Disease

The immune system is a major part of the body involved in protecting us from infections and from cancers. Unlike other organs where the cells and tissues stay in one place (the heart or the kidneys) the cells of the immune system move around the body either to monitor for signs of infection or to fight infection or destroy cancer cells.

Many different types of cells and separate organs make up the immune system. The major components are the bone marrow and thymus (a small organ in the neck) which produce many immune system cells and the spleen (in the abdomen) and the lymph nodes (scattered throughout the body – these are the so called "glands" that swell in the neck or elsewhere when you have an infection) that play a major role in "programming" parts of the immune system.

The immune system is divided into two major parts called the innate and the adaptive.

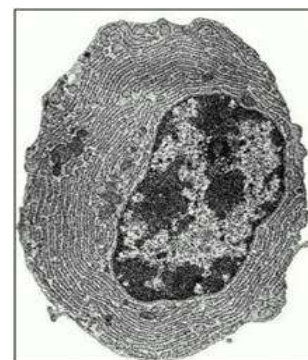
**The Innate immune system** reacts to infections and cancer cells in a pre-programmed way by recognising signals that activate the immune cells to destroy cells or infections with warning signs on them. Neutrophils and macrophages and natural killer cells are the main parts of the innate immune system.



◀ **A highly magnified picture of a neutrophil.**  
**The black dots throughout the cell are antimicrobial granules**

**The Adaptive immune system** is the part of the immune system that can learn to recognise almost any different infection and has to be programmed by the body. This part of the immune system is constantly changing throughout our lives as we encounter new infections, learn how to deal with them and remember them for the future. When we use vaccines we are programming the adaptive immune system to recognise a possible future infection. The adaptive immune system can recognise almost any possible infection by constantly making new cells that are all different. On each cell is a receptor that can match any possible protein or cell component from an infection (in the way a key fits a lock) and when the cell finds its match it can become activated to start making antibodies (if it is a B cell) or making new cells that will direct and control the immune system (if it is a T cell). Unfortunately because the cells can recognise almost anything they may also become activated by parts of our own bodies (this is called autoimmunity). The immune system contains many very specialised systems to prevent cells that recognise our own bodies from becoming activated but sometimes these systems may not work properly and this can lead to autoimmune disease.

**A highly magnified picture of an antibody-producing B cell ►**



Once the immune system has become activated there are many different substances produced by both the immune system and the cells in the place where the infection is. These control the responses by activating cells and summoning more immune system cells to the site of the problem. Some of these substances are proteins called cytokines or chemokines and some treatments for inflammatory diseases block their action to reduce the activation of the immune system.

Unfortunately most treatments for autoimmune diseases and vasculitis cannot specifically target the parts of the immune system causing the problem. Most treatments have a general effect of reducing the activation and effectiveness of the immune system to a greater or lesser extent. This accounts for the main serious short and long term effects of treatment of an increased susceptibility to infections and the small long term increased risk of cancer associated with some treatments.

Usually when doctors are considering the most appropriate treatment for a particular patient they will be trying to get the right balance between suppressing the immune system enough to stop the disease causing damage and long term problems but not suppressing it so much that the risk of infections and cancer is too high. Often the right balance changes over time as the disease comes under control and less strong treatment can be used to keep control of the disease without causing so much suppression of the immune system function.

## **Aetiology (Causes)**

Current research suggests that people probably develop vasculitis because of the complex interaction of their genetic inheritance, which may increase the risk of developing vasculitis, and exposure to chemicals in the environment or possibly some types of infection (including hepatitis B virus) which may trigger the vasculitis in someone who is susceptible. This does not mean that vasculitis can be inherited or passed on to children. The immune system is controlled by many thousands of different genes and it is probable that there needs to be some variation in several different genes in combination to make an individual more "at risk" of vasculitis than others. This does not mean that the genes do not work just that they may work slightly differently. For example, a red apple and a green apple are both apples but they are slightly different. In the same way a single gene may have slightly different variations in different people but still be the same gene doing pretty much the same job. Several genes have been identified where one variety is more common in people with vasculitis and other types of autoimmune disease. This does not mean that the gene caused the vasculitis as many people with the "vasculitis" variety of gene never get vasculitis and some people with the "not vasculitis" version of the gene do get vasculitis. The variety of the gene may not even be specific to one type of vasculitis but may be affecting the way the immune system responds to infections, chemicals or toxins.

## Symptoms

The symptoms caused by vasculitis will depend on the organs involved. However, some general symptoms include: tiredness, weakness, loss of appetite, weight loss, and fever. Unfortunately many of these symptoms apply to other diseases which make it difficult to diagnose vasculitis.

Some symptoms encountered, specific to various systems are:

Respiratory system - breathlessness, wheeze, dry cough or coughing up blood,

ENT (Ear, Nose and Throat) - hearing problems (deafness and or noises in the ear), nasal crusting, nose bleeds, sinus pain (which may be felt as headaches or pain in the face) or hoarse voice,

Skin - rashes, ulcers, and necrosis (death of tissue),

Eyes - red (blood shot) eyes, painful, dry or gritty eyes, visual loss or other changes in vision,

Joints - arthralgias (pain in joints), and joint swelling,

Nervous system - loss of sensation, weakness, unusual painful symptoms in the hands and feet (hotness, pins and needles or "electric shocks") and rarely paralysis or stroke,

Gastrointestinal system - diarrhoea, bleeding and abdominal pain,

Kidneys/Renal - initially no symptoms. However urine dipstick tests will indicate problems with minute amounts of blood or protein in the urine. Occasionally blood may be seen in the urine (red or brown urine) or the amount of urine produced may suddenly reduce or stop altogether.

## Diagnosing Vasculitis

It is essential to obtain a diagnosis of vasculitis as early as possible to enable appropriate treatment. Any delay may result in further permanent damage. The methods for diagnosing vasculitis vary depending on the disease concerned - see individual diseases. However, it is important to realise that diagnosing vasculitis can be problematic for the physician, in part because most of these diseases are rare. Many patients develop symptoms which may be attributable to many diseases as any organ can be affected. The symptoms are often non-specific and often mimic other more common conditions. Unfortunately because of these difficulties diagnosis can be missed or delayed for some time.

Making the right diagnosis will depend on the patient's symptoms, what the doctor finds when examining the patient and often a combination of blood tests, x-rays (or other scans such as MRI and PET) and often a biopsy (taking a small piece of tissue) from an affected area. There is no single test for any of the types of vasculitis.

Once the diagnosis has been made the doctor should discuss with the patient any treatment that is required. Most patients will require some treatment. In some cases of severe disease treatment needs to be given urgently; occasionally in very mild cases no treatment at all is required.

## Treatment

There are two phases in the treatment of vasculitis - "Remission Induction" therapy (getting the disease under control) and "Remission Maintenance" therapy (keeping the disease under control). Both phases normally involve immunosuppressive drugs.

*Remission Induction therapy* - Usually requires a combination of immunosuppressive drugs to control the inflammation. The drugs given will vary according to the specific disease and the severity of the disease. These drugs are commonly high dose steroids (prednisolone) and additional treatment with drugs such as cyclophosphamide or methotrexate may be given. In some types of vasculitis newer antibody treatments (eg rituximab or infliximab sometimes called "biologic therapies") are starting to replace the older treatments such as cyclophosphamide. Usually the amount of steroid treatment will be reduced quickly over the first few weeks and then more slowly. The common side effects of drugs used in remission induction phase are infections because of the suppression of the immune system. Often additional drugs will be given to protect against infection and other side effects of the treatment. (See Glossary of Drugs and Side Effects)

*Remission Maintenance therapy* - Once the disease is controlled or in remission (indicated by improved symptoms and blood tests), the treatment is changed (usually after several months) and maintenance therapy is commenced. The prednisolone is usually reduced to a very low dose. If cyclophosphamide has been given it is usually stopped and changed to less toxic drugs such as azathioprine. Maintenance therapy can last for many years, but in some cases can be discontinued after one or two years. How long people with vasculitis need to remain on remission

maintenance therapy is very variable and will need to be discussed on an individual basis with the medical team helping look after the patient.

Which doctors will be involved in the treatment of vasculitis depends on the type of vasculitis, the organs involved, the severity of the disease and the local practice in your area. You may need several different specialists to manage your disease working as a team. This multidisciplinary team may all be present in one clinic (a multidisciplinary or "one-stop" clinic) or may be working in several clinics in one hospital or even in different hospitals. This may require many different hospital appointments to see the different specialists and good communications between the different members of the team. Information about multidisciplinary clinics may be available from the GP, local specialists or Vasculitis UK.

The treatment given to an individual patient will depend on the type of vasculitis, the severity of the vasculitis, other medical problems the patient may have, how well they have responded to treatment so far and the evidence for the current best treatment available (see section below on clinical trials and research). This means that patients who superficially may appear to have similar disease may get significantly different treatments.

## **Follow-up**

There is no cure for many of the vasculitis diseases. The aim of treatment is to keep the disease process in remission. It is, therefore, essential that the patient is monitored periodically by blood/urine tests and outpatient appointments even when activity of disease is not present.

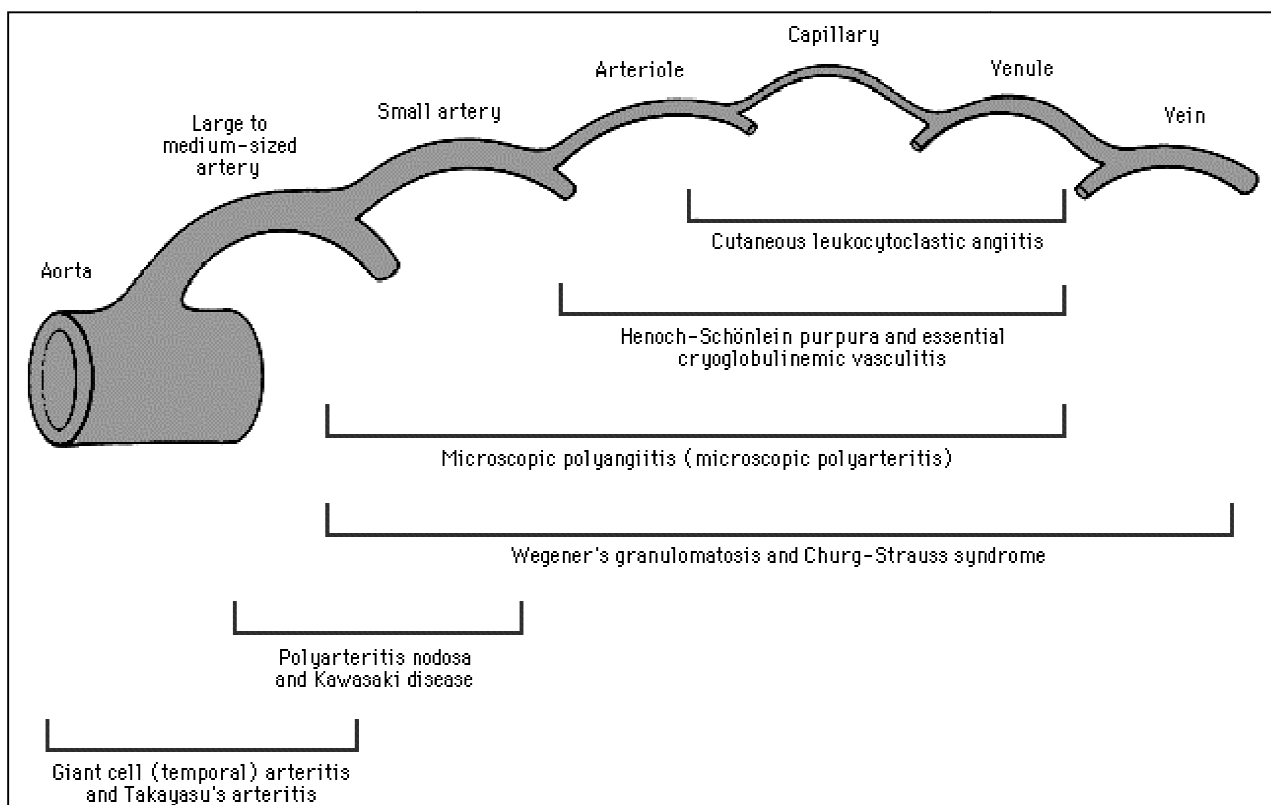
## **Taking part in research and clinical trials**

Doctors treating patients with vasculitis rely on clinical trials and research being conducted and published to know what the best treatments are for different types of vasculitis. This means that the recommendations for treating some types of disease keep changing as they understand more about them and how to treat them. Unfortunately most types of vasculitis are rare and this makes it very difficult to do useful research and conduct clinical trials to find the best treatments. Fortunately there are groups of doctors around the world interested in finding better treatments for vasculitis and by co-operating together research gets done and treatments become safer and more effective.

If you are being treated in a clinic that is involved in vasculitis research and clinical trials you may be invited to take part. If so the medical team should explain the nature of the research or clinical trial to you, give you information sheets to look at, explain the possible pros and cons for you as a patient in taking part, give you the opportunity to ask questions you might have (and give you answers) and respect your decision about taking part whether you choose to participate or not. Sometimes you may benefit directly from taking part in a clinical trial or research, sometimes only other patients in the future will benefit from you having agreed to take part.

## Vasculitis diseases (by size of arteries affected)

As the vasculitic diseases are usually classified according to the predominant size of vessel involved: Detailed information on the individual diseases will be found in the "Individual Vasculitic Diseases" section.



### Vasculitis mainly affecting large arteries

Giant Cell Arteritis/Temporal Arteritis (GCA)  
Polymyalgia Rheumatica (PMR)  
Takayasu Arteritis (TA)

### Vasculitis mainly affecting middle sized arteries

Polyarteritis Nodosa (PAN)  
Kawasaki Disease (KD)

### Diseases affecting the small blood vessels

Also see section below "***Diseases affecting the small blood vessels***"

These are divided into ANCA associated vasculitis and non-ANCA vasculitis.

#### ANCA associated vasculitis

Churg Strauss Syndrome (CSS) now renamed: Eosinophilic Granulomatosis with Polyangiitis (EGPA)  
Microscopic Polyangiitis (MPA)  
Wegener's Granulomatosis (WG) now renamed: Granulomatosis with Polyangiitis (GPA)

#### Non-ANCA vasculitis

Henoch-Schönlein Purpura (HSP)



### **Less common vasculitides**

Behçet's Disease (BD)

Buerger's Disease

Central Nervous System Vasculitis (CNS)/Primary Angiitis of the Central Nervous System (PACNS)

Cogan's Syndrome

Cryoglobulinemia and Cryoglobulinaemic Vasculitis

Hypersensitivity Vasculitis (HSV)

Rheumatoid Arthritis (RA) (secondary vasculitis)

### **Further reading**

**Large Vessel Diseases** - Ashima Gulati, Arvind Bagga

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467\\_2009\\_Article\\_1312.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467_2009_Article_1312.pdf)

**Medium-size Vessel Diseases** - Michael J Dillon, Despina Eletheriou, Paul A Brogan

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467\\_2009\\_Article\\_1336.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467_2009_Article_1336.pdf)

**Small Vessel Vasculitis** - Paul Brogan, Despina Eletheriou, Michael Dillon

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855433/pdf/467\\_2009\\_Article\\_1317.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855433/pdf/467_2009_Article_1317.pdf)

**Small vessel and medium vessel vasculitis** - Philip Seo, John H Stone

<http://onlinelibrary.wiley.com/doi/10.1002/art.23105/pdf>

**Management of ANCA-associated vasculitis: Current trends and future prospects** - Sally Hamour, Alan D Salama, Charles D Pusey

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2893757/pdf/tcrm-6-253.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2893757/pdf/tcrm-6-253.pdf)

## The Individual Diseases

### Giant Cell Arteritis/Temporal Arteritis

#### What is Giant Cell Arteritis/Temporal Arteritis?

GCA is a disease characterised by inflammation of large and medium sized blood vessels. An alternative name for this condition is "Temporal Arteritis" as the blood vessels in the temple area of the head (sides of the forehead) are commonly affected. The giant cells referred to are specific collections of immune system cells seen in the areas of inflammation if a biopsy is performed.

#### Who are affected?

This condition is one of the commonest forms of vasculitis, affecting one or two persons per 10,000 people in the UK. It is more commonly seen in older patients and is seldom diagnosed below the age of 50.

#### What is the aetiology (cause)?

The cause of Giant Cell Arteritis/Temporal Arteritis is not yet known.

#### What are the symptoms?

Symptoms of fatigue, loss of appetite, weight loss and fever are often found. Headache, with pain and tenderness over the temples, is a prominent feature of this disease due to inflammation of the temporal arteries. GCA is closely associated with the "Polymyalgia Rheumatica Syndrome" which causes general weakness, pain and stiffness in muscles and joints. The weakness in the muscles can be quite debilitating and typically affects the upper arms and legs. Simple tasks such as brushing the hair, cooking, gardening, getting up and out of chairs or beds and walking may become difficult.

As this condition affects the main blood vessels supplying parts of the body, a reduction in the blood and oxygen supply (ischaemia) to different organs can result. This can lead to a variety of symptoms. A classic symptom of this condition is pain in the tongue or jaw when eating (jaw claudication). Reduction of the blood supply to the brain can cause a stroke. Reduction in the blood supply to the eyes can lead to blurred vision, or in some patients, blindness.

#### Swollen temporal artery in GCA

Image courtesy of Dr Richard A Watts, University of East Anglia



#### Making a diagnosis

As with most types of vasculitis there is no one test that will give the diagnosis. The diagnosis is usually made because the doctor recognises the typical collection of symptoms and clinical signs. Blood tests for inflammation (ESR – erythrocyte sedimentation rate; CRP – C-reactive protein; plasma viscosity) will usually be high indicating inflammation but these are not specific and can be raised in any type of vasculitis, many infections and other types of inflammatory disease. If the temporal arteries are involved (located at the sides of the forehead over the temples) then a biopsy may be performed. If the biopsy shows inflammation this will help to confirm the diagnosis. Unfortunately a biopsy without inflammation does not exclude the diagnosis. Sometimes an ultra sound scan of the arteries is used instead of a biopsy.

#### Treatment

The two aims of treatment are to relieve patient's symptoms and prevent damage to organs (especially the eyes) due to reduced blood supply. High dose oral steroids (Prednisolone) are usually given and the dose is reduced slowly over months, using the ESR marker as a guide to response. Treatment with steroids is usually given for at least one or two years.

In some patients with resistant or problematic disease other drugs are sometimes used such as methotrexate, Azathioprine or antibody therapy such as infliximab. While some patients may respond well to these drugs there is no current evidence that they are effective in all patients.

### **Side effects**

For the side effects of the drugs used see "Glossary of drugs and side effects".

### **Prognosis**

There is a relapse rate of about 50 per cent. Withdrawal of the steroids can lead to a relapse of the symptoms. In such cases the steroid is re-started and it is often sufficient to induce remission. The steroid dose can again be reduced. In most relapsed cases the patient will be maintained on a small maintenance dose of steroids indefinitely. The prognosis is good with correct and early diagnosis and treatment.

### **Key Points**

- GCA is one of the commonest forms of vasculitis
- Temporal artery biopsy or ultrasound scan may help confirm the diagnosis
- Damage to the eyes is usually the most serious complication
- Treatment is usually with steroids and may continue for many years.

### **Further reading**

**Giant Cell Arteritis: Suspect it, treat it, treat it properly** - Alexandra Villa-Forte  
[www.ccjm.org/content/78/4/265.full.pdf+html](http://www.ccjm.org/content/78/4/265.full.pdf+html)

**The Treatment of Giant Cell Arteritis** - J. Alexander Fraser, Cornelia M. Weyand, et al  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC3014829/pdf/nihms-239316.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3014829/pdf/nihms-239316.pdf)

**Large Vessel Diseases** - Ashima Gulati, Arvind Bagga  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467\\_2009\\_Article\\_1312.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467_2009_Article_1312.pdf)

# Polymyalgia Rheumatica

## What is Polymyalgia Rheumatica?

Polymyalgia Rheumatica (PMR) is an inflammatory disorder. It is frequently linked to Giant Cell Arteritis (GCA) occurring in 50 per cent of patients with GCA. Approximately 15 per cent of PMR patients develop GCA.

## Who are affected?

The disease is usually diagnosed in patients over the age of 65. It is rarely seen in patients younger than 50. Women are twice as likely to develop PMR as men.

## What is the aetiology (cause)?

The cause of PMR is not yet known.

## What are the symptoms?

The symptoms usually come on over a few days. Symptoms include aching muscles and morning stiffness in the shoulders, hips, neck and mid-body. Other symptoms reported include: general tiredness, weakness, weight loss, low grade fever. Inflammation of the bones and joints can cause difficulty moving. Some patients develop swelling or fluid retention (oedema) of the hands, wrists, ankles and top of the feet.

### Illustrations showing normal shoulder joint and inflammation of shoulder joint



Normal bursa sac



Inflammation of bursa



## Making a diagnosis

There are no specific tests for PMR. Blood tests may show evidence of inflammation. Ultrasound and Positron emission tomography (PET scanning) have been used to confirm PMR inflammation. Patients with only PMR do not have the symptoms of GCA therefore a biopsy of the temporal artery (necessary in GCA) is unnecessary in PMR.

## Treatment

Moderate to high doses of steroid drugs (prednisolone) improve the symptoms of PMR. The effective dose is maintained for several weeks after the symptoms have resolved. Then it is gradually lowered and stopped. Careful monitoring is required in case the symptoms recur.

Where long term use of steroids or side effects occur the use of Methotrexate may allow the dose of steroid to be reduced. Ibuprofen (an anti-inflammatory drug) can be used to reduce painful symptoms, especially when the symptoms are only mild.

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

## Prognosis

Relapse occurs in 25 to 50 per cent of patients. This is more likely if the steroid dose is reduced too quickly. Relapse requires restarting or increasing the steroid dose. In most cases the symptoms will cease within one month to one year. The steroids can then be discontinued.

## Key Points

- PMR is sometimes associated with Giant Cell Arteritis
- Treatment is usually with steroids
- Treatment may be able to be stopped once the disease is controlled.

## Further reading

**Treatment of Polymyalgia Rheumatica: A Systematic Review** - José Hernández-Rodríguez, MD, PhD; Maria C. Cid, MD, PhD; Alfons López-Soto, MD, et al  
<http://archinte.jamanetwork.com/searchresults.aspx?q=Polymyalgia&t=&p=1&s=1&c=0>

**Clinical, radiological, and biochemical characteristics in patients with diseases mimicking polymyalgia rheumatica** - Hidekatsu Yanai, Hiroshi Yoshida, and Norio Tada  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2762363/pdf/cia-4-391.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2762363/pdf/cia-4-391.pdf)

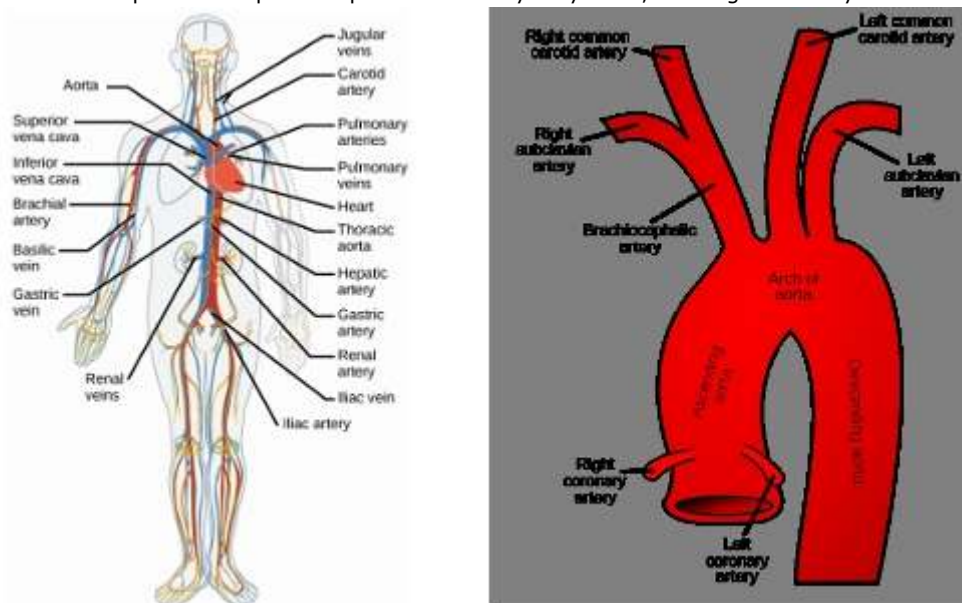
**Large Vessel Diseases** - Ashima Gulati, Arvind Bagga  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467\\_2009\\_Article\\_1312.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467_2009_Article_1312.pdf)

# Takayasu Arteritis

## What is Takayasu Arteritis?

Takayasu Arteritis (TA) is an inflammatory disease of the large arteries. TA particularly affects the aorta (the main artery taking blood from the heart to the rest of the body), and the pulmonary artery (which sends blood to the lungs). The major arteries that arise from the aorta may also be affected. These include the subclavian arteries that supply the arms, renal arteries to the kidneys, coronary arteries in the heart and carotid arteries in the head and brain. (see illustrations below)

In some patients, widening of the aorta results in failure of the aortic valve in the heart, necessitating replacement. In 90% of patients one or more of these arteries become narrowed or blocked, hence TA is also known as "Pulseless disease". In 25 per cent of patients part of an artery may swell, forming an aneurysm.



Illustrations above: Circulation diagram sourced from the "Arteries, Veins, and Capillaries" section of Robert Bear and David Rintoul's article, "The Circulatory System," Connexions, August 19, 2013, The Circulatory System. Aorta - Ascending, Arch & Descending diagram sourced from: "Gray's Anatomy" Wikipedia Commons, Aorta diagram.

## Who are affected?

TA is a very rare disease of young people with the initial symptoms arising in those between 5 and 40 years of age. Although both sexes may be affected, 80-90 per cent of patients are female. TA has worldwide prevalence and may affect all races. However, the disease is thought to be more common in patients originating from the Far East, Japan and the Asian sub-continent. As far as we know, TA is not a genetically inherited disease. It is unusual for more than one family member to suffer from the disease. However, the increased prevalence in certain parts of the world may indicate a genetic component to disease susceptibility and current studies are investigating this.

## What is the aetiology (cause)?

The cause of Takayasu's arteritis, and why an individual develops the disease at any one time remains unknown. The disease is most likely to be the consequence of environmental factors and a susceptible genetic background. One common but unproven hypothesis is that it is precipitated by an infection. The rarity of the disease worldwide means that it is very difficult to identify an underlying cause. The inflammation involves white blood cells invading the wall of the artery predisposing to damage and scarring.

## What are the symptoms?

The initial symptoms of TA are typically non-specific and may include one or more of:- malaise (a general feeling of being unwell), profound fatigue, fever, night sweats, weight loss, myalgia (muscle pain), arthralgia (painful joints), rash. Additional symptoms can include dizziness/light headedness, shortness of breath, cramping pain in the arms, legs or chest on exertion. Carotidynia, (pain and tenderness over the carotid arteries in the front of the neck) is also found in approximately 25 per cent of patients.

## Making a diagnosis

This is made on the basis of formal criteria such as those published by the American College of Rheumatology. For diagnosis the patient should fulfil three or more of the criteria listed below:

### Diagnostic Criteria for Takayasu's Arteritis

1. Onset of disease  $\leq$  40 years
2. Claudication\*\* of an extremity
3. Reduced brachial artery pulsation
4. Difference in systolic blood pressure  $>10$  mmHg between the arms
5. Aortic or subclavian artery bruit
6. Angiographic abnormality

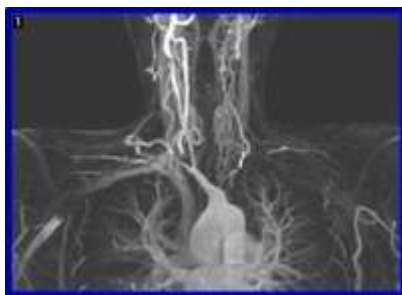
\*\* Claudication - severe muscle pain due to cramp, caused by narrowed blood vessels

In practice clinical examination most commonly detects decreased or absent pulses in the arms and less frequently in the legs. Using a stethoscope, bruits\*, (a loud 'whooshing noise') may be detected, over the neck, chest or kidneys indicating narrowed arteries. High blood pressure is commonly found. In recent years specialist scans have been introduced to aid diagnosis and identify inflammation, narrowing and dilatation of arteries and the extent of the disease. These include positron emission tomography (PET scanning), magnetic resonance angiography (MRA), high-resolution ultrasound and CT angiography (see below).

### Diagnostic tests

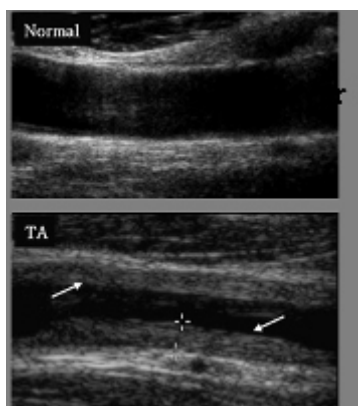
**Blood Tests:** There is no blood test that can be used to definitively confirm the diagnosis. The erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) can help measure activity of the illness and your response to treatment.

**Imaging:** Imaging studies are useful in the diagnosis and monitoring of TA. Their use may allow diagnosis before arterial damage is severe. However, none of them are 100 per cent accurate.



**Magnetic resonance imaging (MRI)** is commonly used to aid diagnosis and assess the extent of TA. The scan does not involve radiation and the picture of the arteries can be enhanced by using a contrast medium (that only rarely causes an allergic reaction).

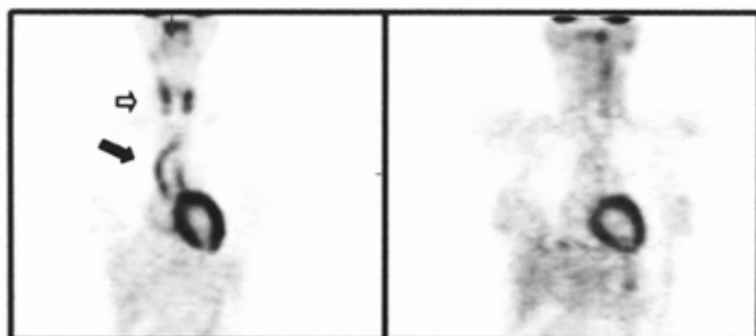
MRI is a safe means by which to monitor the progress of disease and response to treatment. The main drawbacks are that the scanner is very noisy and rather claustrophobic. The illustration shows an MRI scan of a TA patient.



**Ultrasound - Doppler ultrasound** (ultrasound scan): uses a probe which is passed over the carotid arteries in the neck. This allows assessment of blood flow and can detect and monitor the arterial wall and the degree of narrowing.

This illustration shows a normal carotid artery and one from a TA patient. The arrows show the thickening of the artery wall causing the narrowing of the artery.

**Positron emission tomography (PET scanning):** requires the patient to be injected with radioactive fluorodeoxyglucose. PET scans are particularly good at detecting inflammation in the arterial wall and are often used in combination with CT scanning (CT-PET). PET may identify early active disease and may on occasion help to distinguish active disease (which needs further treatment) from scarring resulting from previous inflammation. The illustration shows two PET images from the same patient; the one on the left is before treatment, with the arrows pointing to areas of inflammation. These areas have disappeared in the image on the right which was taken after six months treatment.



**Computed tomography (CT) angiography:** Contrast-enhanced CT angiography is useful for the diagnosis of TA and can help identify the extent of damage to the arteries. Unlike MRI/MRA and Doppler US, CT angiography does involve radiation.

### Treatment

Treatment is aimed at improving symptoms and preventing further damage and/or scarring to the blood vessels. When there is active inflammation in the arteries, treatment is started with steroids, usually prednisolone, and often in combination with an immunosuppressant drugs. Those commonly used include methotrexate, azathioprine or mycophenolate. These drugs increase the effectiveness of steroid therapy and help to reduce the dose of steroids required. In patients with persistent or very severe disease, cyclophosphamide may be recommended. In recent years, for refractory disease, biologic therapies, including tumour necrosis factor inhibitors such as etanercept and infliximab have been used successfully, as has tocilizumab which targets interleukin-6.

**Additional treatment:** Drugs to lower blood pressure are often recommended. Statins, the most common of which is simvastatin, are commonly prescribed to lower cholesterol and reduce the risk of heart disease. In TA they may also have a beneficial effect on arterial function, even when cholesterol levels are normal. Low-dose aspirin may be prescribed to reduce the risk of a blood clot developing.



**Surgery:** Surgical intervention should not be undertaken lightly and is only indicated for the most severe cases of TA. For optimal results surgery should be carried out when the disease is in remission and decisions will involve a team of specialist doctors including the rheumatologist, vascular surgeons and radiologists. Procedures that may be carried out include:

**Revascularisation** - where a blocked artery is bypassed, typically with a vein graft from somewhere else in your body. Good long-term outcomes have been seen, with 20 year graft survival rates >70 per cent in one study. However, results are variable and further studies are needed for more conclusive evidence.

**Percutaneous transluminal angioplasty (PTA)** - aims to open narrowed arteries by inserting and inflating a balloon to stretch the narrowed section. In some cases a wire mesh (stent) may be required to keep the artery open.

### Drugs and Side effects

For information on the main drugs prescribed for Takayasu Arteritis see:

- Azathioprine
- Cyclophosphamide
- Methotrexate
- Mycophenolate Mofetil (Cellcept)
- Steroids

Information on drugs and side effects can be found at: [www.vasculitis.org.uk/about-vasculitis/glossary-of-drugs](http://www.vasculitis.org.uk/about-vasculitis/glossary-of-drugs)

### Pregnancy and Takayasu arteritis

Many women with Takayasu's arteritis achieve one or more successful pregnancies.

**It is very important for a woman planning to have a baby to talk to her specialist before trying to conceive.** Depending on how advanced your disease is and how much damage has been done to your arteries, it may on occasion be dangerous for a woman to carry a baby. For example control of blood pressure can be difficult in patients with TA and blood pressure can be increased by pregnancy to a dangerous level. Likewise, high pressure in the pulmonary (lung) circulation can place a woman at significant risk during pregnancy.

Unless the disease is very active, TA itself will not affect your fertility, whether you are a man or a woman. However, some of the drugs used to treat the condition may reduce fertility, Likewise, other medications may increase the risk of defects in the foetus. It is therefore essential for both women and men with TA to consult their specialist to discuss all aspects of starting a family.

### Prognosis

The long term prognosis of TA is good. Approximately 20 per cent of patients will have a monophasic self-limiting disease (just one inflammatory episode). More typically, the disease follows a relapsing and remitting course. Limited data is available on long-term outcome and varies significantly between patients, dependent upon the extent of their disease. In the majority of patients the TA appears to 'burn out' after a period of 2-5 years, and treatment can be gradually withdrawn. The figures below summarise data from surveys performed around the world:

- 25 per cent of patients largely unaffected by their disease
- 25 per cent of patients able to perform normal daily activities when disease in remission
- 50 per cent were unable to consistently do or maintain their full range of daily activities
- Up to 25 per cent of patients are unable to work due to their Takayasu's arteritis

With early intervention and careful monitoring in a specialist centre TA can be effectively treated. Mortality rates are low with survival in the USA and UK up to 98 per cent at 10 years. Mortality directly related to TA is usually the consequence of heart attack, heart failure, aneurysm rupture, stroke or kidney failure.

### Key Points

- TA is very rare in the UK and mainly affects young women
- Presenting symptoms are often non-specific and missed

- Treatment with steroids and immunosuppressants is effective in many cases
- Surgical repair or angioplasty to narrowed blood vessels should only be performed in severe cases and after the inflammation is controlled

### **Acknowledgements**

Vasculitis UK is indebted to Prof Justin Mason of Imperial College, London, for producing this web page on Takayasu Arteritis.

### **Related Vasculitis Articles**

- Fertility and Vasculitis - Dr David Jayne - [www.vasculitis.org.uk/living-with-vasculitis/fertility](http://www.vasculitis.org.uk/living-with-vasculitis/fertility)
- 
- Vasculitis in Children - Dr Paul Brogan - [www.vasculitis.org.uk/living-with-vasculitis/vasculitis-in-children](http://www.vasculitis.org.uk/living-with-vasculitis/vasculitis-in-children)

### **Further reading**

- Large Vessel Diseases - Ashima Gulati, Arvind Bagga  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467\\_2009\\_Article\\_1312.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467_2009_Article_1312.pdf)
- Vasculitis Paediatric Guidelines 2012 - Dr Helen Foster & Dr Paul Brogan, Oxford University Press - [www.vasculitis.org.uk/professionals/paediatric-vasculitis](http://www.vasculitis.org.uk/professionals/paediatric-vasculitis)

## Polyarteritis Nodosa

### What is Polyarteritis Nodosa?

Polyarteritis Nodosa (PAN) is a very rare vasculitic disease which affects the medium sized vessels. It can affect any organ in the body but commonly the muscles, joints, intestines, nerves and skin are affected. It can be associated with Hepatitis B virus infection.

### Who are affected?

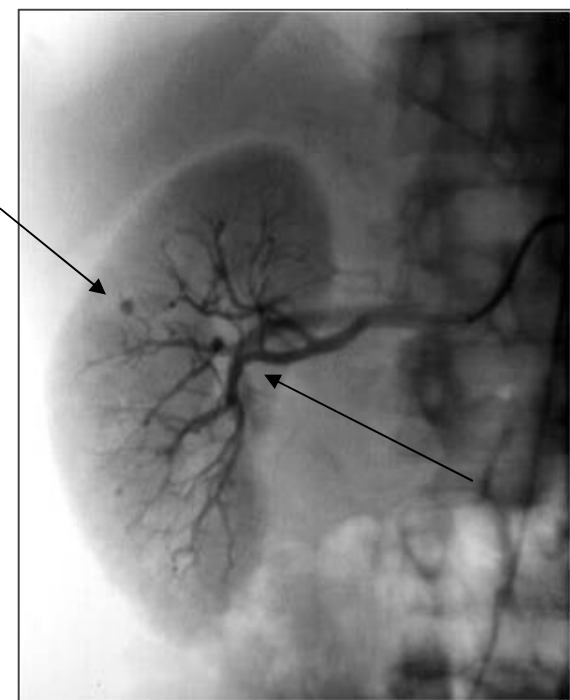
PAN is most common in the middle-aged.

### What is the aetiology (causes)?

The cause of PAN is not yet known. In some people there is a clear association between PAN and hepatitis B virus infection although why this is the case is not entirely clear. Only a very few people with hepatitis B infection ever develop PAN and only a small proportion of people with PAN have hepatitis B infection.

### What are the symptoms?

It depends on the area affected. If the bowels are affected this can lead to pain in the abdomen or severe pain after eating (mesenteric angina). Sometimes the reduction of blood flow to the bowels leads to inflammation, bleeding, ulcers and sometimes perforation of the bowel. Involvement of the nerves can lead to numbness, pain or pins and needles in the affected nerves and weakness in the muscles. Aching in the muscles and joints is common and some patients develop skin rashes or ulcers.



### ◀ A kidney angiogram in a patient with PAN

The aneurysms (bubbles in the wall of arteries) are indicated by the arrows.

Image courtesy of Dr David Jayne, Addenbrooke's, Cambridge, UK

### Diagnosis

As with other types of vasculitis there is no one diagnostic test. Diagnosis is based on the symptoms described by the patient, physical examination, laboratory tests and possibly biopsy of the affected area. Blood tests will show evidence of inflammation. The blood vessels in the abdomen are commonly affected and angiography (x-rays of the blood vessels) can show typical findings which help with the diagnosis.

It is important to test patients with PAN for hepatitis B virus as this may require separate treatment.

### Treatment

High dose steroids and cyclophosphamide are often used to get the inflammation under control (remission induction). Once under control treatment is often continued for 12 months with low dose treatment and then can be stopped in some patients.

Patients with hepatitis B virus infection will need specific treatment for the virus infection with anti-viral drugs, usually under specialist care.

### **Side effects**

For the side effects of the drugs used see "Glossary of drugs and side effects".

### **Prognosis**

With treatment the prognosis is generally good. In most cases treatment can be stopped after 12 months. In some patients the disease will relapse requiring longer term treatment.

### **Key Points**

- PAN is a rare type of vasculitis
- It may be associated with hepatitis B virus infection
- Treatment depends on the severity and the presence of hepatitis B virus.

### **Further reading**

**Medium-size Vessel Diseases** - Michael J Dillon, Despina Eletheriou, Paul A Brogan  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467\\_2009\\_Article\\_1336.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467_2009_Article_1336.pdf)

# Kawasaki Disease

## What is Kawasaki Disease?

Kawasaki Disease (KD) (also known as Kawasaki Syndrome) is a rare form of vasculitis which affects children and is characterised by inflammation of the medium sized blood vessels throughout the body. The coronary arteries (arteries supplying the heart muscle) are the commonest arteries affected. The main concern in children is that inflammation of the coronary arteries can lead to aneurysms (widening and weakening of the artery wall) and later can lead to narrowing or stenosis reducing the blood supply to the heart.

## Who are affected?

Primarily this disease affects young children under the age of 5 years. Older children and teenagers can develop KD but this is rare. The disease is more common in boys than girls.

## What is the aetiology (cause)?

The cause of KD is not yet known. Some studies in KD suggest that infection may play an important role in triggering the disease as the incidence of the infection varies with the season of the year. As yet no definite link to a particular infection has been proven.

## What are the symptoms?

Fever (lasting for five or more days), rash (worse in the groin area), bloodshot eyes, red, swollen, cracked lips. "strawberry" tongue with shiny bright red spots, swollen hands and feet with redness of the palms and soles of the feet, swollen lymph nodes in the neck.

## Making a diagnosis

There are no specific diagnostic tests for KD. Clinicians diagnose the condition based on physical examination and history. Blood tests will show evidence of inflammation.

## Treatment

Early diagnosis is essential and treatment is by a combination of intravenous gamma-globulin (which is normal antibodies taken from blood donors) and aspirin. A few patients may also require additional treatment with other immunosuppressants.

Some children may develop long-term coronary artery problems which require long-term care under a cardiologist.

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

## Prognosis

In the majority of patients the disease is self limiting. Treatment aims to reduce problems with the coronary arteries which can lead to long term problems.

## Key Points

- KD usually only affects young children
- Damage to the coronary arteries is the commonest serious problem of KD
- Treatment does not usually need immunosuppressants.

## Further reading

**Pathogenesis and management of Kawasaki disease** - Anne H Rowley, Stanford T Shulman  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2845298/pdf/nihms180284.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2845298/pdf/nihms180284.pdf)

**Medium-size Vessel Diseases** - Michael J Dillon, Despina Eletheriou, Paul A Brogan  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467\\_2009\\_Article\\_1336.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467_2009_Article_1336.pdf)

## Diseases affecting the small blood vessels

The diseases that affect the small vessels include some of the most common types of vasculitis. They are often divided into the diseases where anti-neutrophil cytoplasm antibodies (ANCA) are found in the blood (ANCA associated vasculitis) and diseases where ANCA is not found. The ANCA associated vasculitides are Wegener's granulomatosis (now being called Granulomatosis with Polyangiitis, Microscopic Polyangiitis and Churg-Strauss syndrome (now being called Eosinophilic Granulomatosis with Polyangiitis).

Many of the small vessel diseases can cause damage to the kidneys and prevent them working. The typical symptoms of impaired kidney function would be fatigue, poor appetite, and swollen legs and breathlessness due to retention of fluid. In severe cases acid and toxins accumulate in the body causing severe breathlessness, nausea and vomiting, confusion and epileptic fits. The blood pressure may also be very high which can cause damage to the kidneys, heart, brain and other organs.

Any patient with vasculitis should have the urine tested for blood and protein using a dipstick test. The presence of blood and protein can indicate inflammation in the kidneys and requires urgent assessment (within a few days) by a kidney specialist and may need a kidney biopsy. If not treated urgently inflammation in the kidneys can cause permanent damage needing dialysis treatment or kidney transplantation.

In the early stages of kidney inflammation the blood tests of kidney function (creatinine and estimated glomerular filtration rate (eGFR)) may still be normal – this is the best time to treat before significant damage has occurred.

## Churg Strauss Syndrome (now renamed Eosinophilic Granulomatosis with Polyangiitis (EGPA))



Jacob Churg



Lotte Strauss

and

### Change of nomenclature

The name of this vasculitic disease was officially changed from Churg Strauss Syndrome (CSS) to Eosinophilic Granulomatosis with Polyangiitis (EGPA) in 2011. Throughout this descriptive section it will be referred to as EGPA.

### What is Eosinophilic Granulomatosis with Polyangiitis?

EGPA was first described by Drs Churg and Strauss in 1951. It is also known as “allergic granulomatous angiitis”. It is usually diagnosed in patients with the combination of asthma (which may have only recently started), an increased eosinophil count (a type of white blood cell) in the blood and vasculitis (inflammation) of the blood vessels.

### Who are affected?

EGPA predominantly affects patients aged 30-45, either male or female, the highest rate being in males. It is uncommon in those over 65 and in children.

### What is the aetiology (cause)?

The cause of EPGA is not yet known.

### What are the symptoms?

Many of the symptoms are non-specific and include fever, muscle and joint aches, tiredness, loss of appetite and weight loss.

It is common for the lungs to be affected in EGPA either by asthma or sometimes by more direct inflammation of the blood vessels causing breathlessness.

Inflammation of vessels supplying nerves in the arms and legs is common and more than 60 per cent of patients will notice numbness affecting discrete areas of their arms or legs or weakness in the muscles of the arms or legs. Less often, nerves in the head can be affected and patients may notice hearing loss or blurred vision. Less commonly the kidney may be affected causing reduced kidney function and high blood pressure or the heart may be affected leading to heart failure. Sometimes the bowels can be affected leading to pain in the abdomen.



◀ Skin involvement in EGPA  
Image courtesy of an EGPA patient

## Diagnosis

As in other types of vasculitis there is no single test which makes the diagnosis. Blood tests usually show evidence of inflammation. The white cell count in the blood can be abnormal with high numbers of eosinophils seen. The blood tests for ANCA (Anti-neutrophil cytoplasmic antibodies) are present in the blood in 50 per cent of patients.

Biopsy of an organ such as the kidneys or lungs may show blood vessel inflammation and accumulations of eosinophils and other blood cells.

## Treatment

Initial treatment to induce remission is usually with high dose steroids and possibly additional immune system suppressing drugs depending on the severity of the disease. Commonly cyclophosphamide will also be given to patients with severe disease. Once remission is established treatment with low dose steroids and sometimes additional drugs such as azathioprine or methotrexate is continued for the longer term.

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

## Prognosis

EGPA can relapse. The prognosis is good with correct and early diagnosis and treatment. Overall prognosis depends on the disease severity.

## Key Points

- The main features of EGPA are vasculitis and asthma
- The treatment depends on the severity of the disease
- Overall prognosis depends on the disease severity.

## Further reading

**Churg-Strauss Syndrome: Evolving Concepts** - Christian Pagnoux

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## Microscopic Polyangiitis

### What is Microscopic Polyangiitis?

Microscopic Polyangiitis (MPA) is inflammation of the small and sometimes medium sized blood vessels. In the past MPA was thought to be a type of PAN and patients would have been told they had PAN. It is now recognised that they are separate diseases.

In microscopic polyangiitis it is common for the smallest blood vessels (capillaries) of the lungs and kidneys to be damaged leading to reduced kidney function or kidney failure and breathlessness.

### Who are affected?

MPA is commonest in middle aged people and affects both men and women.

### What is the aetiology (cause)?

The cause of MPA is not yet known

The ANCA antibodies found in most patients with MPA play a role in causing the inflammation of the blood vessels by activating some types of white blood cell. The ANCA antibodies attach to the neutrophils in the blood causing activation. This makes the neutrophil attach to the blood vessel wall and cause damage by release of the chemicals that it usually uses to fight infection. It is not understood why patients develop ANCA antibodies.

### What are the symptoms?

Common symptoms in MPA are tiredness, loss of appetite and joint and muscle aches. The other symptoms of MPA will depend on the organs involved and will vary markedly between different patients.

Occasionally MPA just affects the kidneys (sometimes called Renal Limited Vasculitis) so that an individual may only begin to feel unwell when the kidney function has deteriorated.

Some symptoms encountered, specific to various systems are:

Lungs - breathlessness, wheeze, dry cough or coughing up blood.

Skin - rashes, ulcers, and necrosis (death of tissue).

Eyes - red (blood shot) eyes, painful, dry or gritty eyes, visual loss or other changes in vision.

Nerves - loss of sensation, weakness, unusual painful symptoms in the hands and feet (hotness, pins and needles or "electric shocks") and rarely paralysis or stroke.

Bowels - Diarrhoea, bleeding and abdominal pain.

### Skin ulcer in MPA ►

Image courtesy of a Vasculitis UK Member



### Diagnosis

As with other type of vasculitis the diagnosis depends on the doctor recognising the pattern of symptoms and clinical signs. The ANCA blood test and a biopsy from the kidney or other affected organ are very helpful tests for MPA.

### Treatment

Prednisolone (steroid) and cyclophosphamide are often used to get the disease under control in moderate or severe cases. In mild cases where the kidneys are normal milder treatment with prednisolone and methotrexate may be given. If the disease is very severe large doses of methylprednisolone or plasmapheresis (plasma exchange) may also be given. When the disease becomes quiet less toxic drugs are used to keep control which include: azathioprine, methotrexate and mycophenolate mofetil usually in combination with low dose prednisolone.

Rituximab has recently been shown to be effective in some cases of MPA instead of cyclophosphamide although it is not suitable for all patients.

### **Side effects**

For the side effects of the drugs used see "Glossary of drugs and side effects".

### **Prognosis**

The overall prognosis in MPA depends on the severity of the disease and the amount of damage that has been done to organs, especially the kidneys, when the disease was active.

Relapses can occur in MPA though less commonly than in some other types of vasculitis and usually require a temporary increase in treatment. It is not clear how long patients with MPA should remain on treatment for and this should be discussed with the doctors looking after the patient on an individual basis.

### **Key Points**

- MPA is a rare form of vasculitis
- It often causes severe kidney and lung damage
- Treatment often requires high dose steroids and cyclophosphamide.

### **Further reading**

**Microscopic Polyangiitis** Sharon A. Chung, Philip Seo

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# Wegener's Granulomatosis (now renamed Granulomatosis with Polyangiitis (GPA))

## Change of nomenclature

The name of this vasculitic disease was officially changed from Wegener's Granulomatosis (WG) to Granulomatosis with Polyangiitis (GPA) in 2011. Throughout this descriptive section it will be referred to as GPA.

## What is Granulomatosis with Polyangiitis?

GPA is a type of primary systemic ANCA associated vasculitis (AAV). It is the most common type of this group of vasculitis diseases. It usually affects the kidneys, lungs, ears, nose and sinuses. GPA is characterised by inflammation of the small blood vessels including the capillaries.

## Who are affected?

It is most common in middle aged and elderly people but can affect young adults and children. It affects men and women equally.

## What is the aetiology (cause)?

The cause of GPA is not yet known. Some research suggests that GPA may be triggered by exposure to silica or to infection with staphylococcus aureus bacteria though this is not proven.

The ANCA antibodies found in most patients with GPA play a role in activating the disease. The ANCA antibodies attach to the neutrophils in the blood causing activation. This makes the neutrophil attach to the blood vessel wall and cause damage by release of the chemicals that it usually uses to fight infection. It is not understood why patients develop ANCA antibodies.

## What are the symptoms?

The disease can present in very different ways in different people depending on the severity and the organs involved. It is not uncommon for patients to have had mild symptoms for months or even years before seeing a doctor at all.

Common general symptoms include tiredness, loss of appetite and aching muscles and joints.

It is very common for GPA to affect the ears, nose and sinuses causing blocked nose with some bleeding, crusts and blood clots. Deafness is also very common due to inflammation in the ears as is pain in face or headaches due to sinus inflammation.



**Episcleritis in GPA**

Image courtesy of Dr David Jayne Adenbrooke's, Cambridge UK

In some patients the kidneys can be severely affected which leads patients to go to a doctor with symptoms of kidney disease.



**Skin rash in GPA**

Images courtesy of Dr Richard A Watts, University of East Anglia



**Scleritis in GPA**

Other common symptoms include:

Lungs - breathlessness, wheeze, dry cough or coughing up blood.

Skin - rashes, ulcers, and necrosis (death of tissue).

Eyes - red (blood shot) eyes, painful, dry or gritty eyes, visual loss or other changes in vision.

Nerves - loss of sensation, weakness, unusual painful symptoms in the hands and feet (hotness, pins and needles or "electric shocks") and rarely paralysis or stroke.

Bowels - Diarrhoea, bleeding and abdominal pain.

#### Saddle nose in a patient with GPA



▲ Pre-operative



▲ After Reconstruction

Images courtesy of a Vasculitis UK Member

#### Diagnosis

As in other types of vasculitis there is no single test which confirms the diagnosis. The diagnosis will depend on the doctor recognising the pattern of symptoms and examination findings. Blood tests usually show evidence of inflammation. The blood tests for ANCA are usually positive which help support the diagnosis but are not specific for GPA or vasculitis. If the kidneys or lungs are affected then a biopsy may be helpful in confirming the diagnosis.

GPA often presents in a similar way to other diseases and can be difficult to diagnose. If the lungs are primarily affected it is not uncommon for the disease to be mistaken for lung cancer or TB.



#### Chest x-ray showing pulmonary haemorrhage in GPA ►

Image courtesy of Dr Richard A Watts, University of East Anglia

#### ◀ Chest x-ray showing nodules in GPA

Image courtesy of Dr Richard A Watts, University of East Anglia



## Treatment

Prednisolone (steroid) and cyclophosphamide are often used to get the disease under control in moderate or severe cases. In mild cases where the kidneys are normal milder treatment with prednisolone and methotrexate may be given. If the disease is very severe large doses of methylprednisolone or plasmapheresis (plasma exchange) may also be given. When the disease becomes quiet less toxic drugs are used to keep control which include: azathioprine, methotrexate and mycophenolate mofetil usually in combination with low dose prednisolone.

Rituximab has recently been shown to be effective in some cases of GPA instead of cyclophosphamide although it is not suitable for all patients.

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

## Prognosis

The overall prognosis in GPA depends on the severity of the disease and the amount of damage that has been done to organs, especially the kidneys, when the disease was active.

Relapses can occur quite commonly in GPA and usually require a temporary increase in treatment. It is not clear how long patients with GPA should remain on treatment for and this should be discussed with the doctors looking after the patient on an individual basis.

## Key Points

- GPA is the most common type of this group of vasculitis diseases
- The disease may be present for months or years before a diagnosis is made
- Treatment depends on the severity of the disease
- The disease commonly relapses after the initial treatment.

## Further reading

**Classification, presentation, and initial treatment of Wegener's granulomatosis in childhood** - David A Cabral, America G Uribe, Susanne Benseler, et al  
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## Photo library - Wegener's Granulomatosis (GPA)



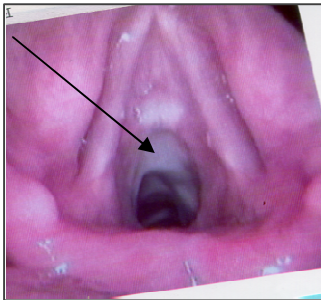
**Swelling and inflammation pre-diagnosis of GPA**

Image courtesy of a Vasculitis UK Member



**Skin lesion in GPA**

Image courtesy of GPA patient



◀ **Subglottic stenosis in GPA (Wegener's).** Note the narrowing to the airway (arrowed) just below the larynx.

Image courtesy of Dr David Jayne, Addenbrooke's, Cambridge, UK

**Untreated GPA (Wegener's) eroding through the skin between the nose and the right eye. ▶**

Image courtesy of Dr David Jayne, Addenbrooke's, Cambridge, UK



◀ **'Strawberry gingivitis'. GPA (Wegener's) causing vasculitis of the gums.**

Image courtesy of Dr David Jayne, Addenbrooke's, Cambridge, UK



◀ **High powered view of exudates on the edge of the cornea associated with episcleritis in GPA.**

Image courtesy of Dr David Jayne, Addenbrooke's Cambridge, UK

**Severe scleritis causing destruction of the cornea in GPA (Wegener's) ▶**

Image courtesy of Dr David Jayne, Addenbrooke's Cambridge, UK



◀ **Collapse in the bridge of the nose caused by GPA (Wegener's).**

Note the hearing aid required for hearing loss due to concurrent middle ear disease

Image courtesy of Dr David Jayne, Addenbrooke's Cambridge, UK

# Henoch Schönlein Purpura

## What is Henoch-Schönlein Purpura?

Henoch-Schönlein Purpura (HSP) is a systemic vasculitis which can affect the skin, joints, bowel and kidneys. If it affects the kidneys it is usually called Henoch-Schönlein Nephritis or Vasculitic IgA Nephropathy.

## Who are affected?

HSP mostly affects children, but can affect adults. Sometimes it follows a throat or chest infection. It affects boys and girls equally. Half the children affected are under the age of five. Kidney involvement is more likely to be severe in older children and adults.

Sometimes HSP is occasionally also called Berger's disease but this should not be confused with **Buerger's** disease which is a different type of vasculitis

## What is the aetiology (cause)?

The cause of HSP is not yet known.

## What are the symptoms?

Symptoms occur over days or several weeks. A widespread rash is common mainly on the backs of the legs, buttocks, trunk and back. The rash is caused by inflammation in the small blood vessels of the skin which can cause bleeding into the skin. It is often felt as small bumps (palpable) and is a reddish/purple colour (purpura). In severe cases it can cause large areas of raised purple patches and skin ulcers. Other common symptoms are painful joints, stomach and abdominal pain and sometimes bleeding from the bowels. If the kidneys are involved there will be blood and protein in the urine.



### ◀ Skin rash in HSP

Image courtesy of Dr Richard A Watts University of East Anglia

## Making a diagnosis

As with other types of vasculitis there is no single specific test. The diagnosis depends on the doctor recognising the pattern of symptoms and findings on examination. Blood tests may show evidence of inflammation. There may be a raised level of immunoglobulin A (IgA) (a type of antibody) in the blood. If the skin or kidneys are affected then a biopsy may show immunoglobulin A antibodies present in the tissue. A positive biopsy helps confirm the diagnosis but a negative biopsy does not rule out the diagnosis.

## Treatment

No specific treatment is needed for most cases of HSP with the symptoms resolving spontaneously over time. Anti-inflammatory drugs (such as ibuprofen) are often prescribed for the joint pains. There is some evidence that steroid treatment may lead to more rapid improvement of bowel symptoms in children. If the kidney is involved and there is inflammation in the kidney then treatment with steroids or other immunosuppressants may be needed to prevent damage to the kidneys.

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".





**Blisters pre-diagnosis in HSP**

Image courtesy of a Vasculitis UK Member



**Skin rash in HSP**

Image courtesy of a Vasculitis UK Member

### **Prognosis**

Although relapses are quite common they are mostly mild and self-limiting. Usually the disease stops re-occurring as patients get older.

### **Key Points**

- The disease may be present for months or years before a diagnosis is made
- Treatment depends on the severity of the disease
- The disease commonly relapses after the initial treatment.

### **Further reading**

**Henoch-Schönlein Purpura** - Brian V Reamy, Pamela M Williams, et al  
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<http://onlinelibrary.wiley.com/doi/10.1002/art.23105/pdf>

# Behçet's Syndrome

## How is Behçet's usually pronounced and why?



Behçet's is usually pronounced as "Beh-CHETS" after Hulusi Behçet a Turkish Professor who spent many years promoting the clinical awareness of the condition.

## Is Behçet's syndrome' and Behçet's disease' the same condition?

The condition has been given many names, with the two most widely accepted being Behçet's syndrome and Behçet's disease. We favour using syndrome over disease as syndrome suggests that not every patient has exactly the same condition, whilst disease implies a single entity.

## What is Behçet's Syndrome?

Behçet's syndrome is an inflammatory condition that may affect many different parts of the body. Often the skin and the lining of the mouth and genital areas (mucosa) can be inflamed. Joint pains, headaches, tiredness and stomach pains are also common. In some people, the eyes, blood vessels or (rarely) the brain or nerves may become involved.

## Who are affected?

Behçet's syndrome is uncommon. It is most common along the ancient silk trading routes stretching from the Far East to the Mediterranean, and appears to have the highest frequency in Turkey and surrounding regions.

Although it is quite rare in the UK and other Northern European countries, it may occur in people of any nationality and race. Men and women are probably affected with equal frequency, but it is sometimes more severe in men. It may occur at any age, but is diagnosed most commonly in adults in their 20s and 30s. It is very unusual in young children.

## What is the aetiology (cause)?

### Genetics

Behçet's syndrome is not well understood by medical professionals. It is likely that people have a genetic predisposition. That means that they have certain genes that make them more likely than others to develop the condition. Doctors think this is the case partly because of the fact that it is more common in some countries than in others, and partly from genetic studies. Having an affected family member very marginally increases your risk of developing the condition. As the syndrome is not present from birth, doctors feel that it is likely that genes aren't the only factor involved. Indeed, if one of a pair of identical twins (who have identical genes) develops Behçet's Syndrome, the other twin will not necessarily develop the condition.

### Environmental Triggers

It is likely that an environment factor(s) trigger is necessary to cause Behçet's syndrome to develop in individuals with a genetic predisposition' (above). Researchers have studied many viruses (e.g. Herpes cold sore virus), bacterial infections (e.g. bugs that cause a 'Strep throat'), smoking, and the bugs that are present in the bowel, but so far have not fully answered this question.

### Resultant abnormal inflammation

The result of the genes interacting with the environmental trigger is that someone who goes on to have Behçet's Syndrome develops abnormal inflammation. Inflammation is a necessary function of the body, being needed to fight infections. In people with Behçet's syndrome, the inflammatory response becomes too enthusiastic, whereby a small insult to the body (e.g. a thorn prick) may result in a big inflammatory response, and inflammation may occur without an obvious cause.

## What are the symptoms?

### Ulcers



Recurrent mouth ulcers are the hallmark of Behçet's syndrome. Almost all patients have experienced these, and they can predate other symptoms by months or even years. Ulcers can be small, large or multiple. They are often on the inner lips but can also occur on the tongue, the roof of the mouth and occasionally the throat. Ulcers in Behçet's syndrome do not typically form on the external lips. The ulcers are usually painful and occasionally leave a scar. Mouth ulcers affect up to 10% of the population so the presence of mouth ulcers alone is not enough for a diagnosis of Behçet's Syndrome. Importantly, many other conditions can also be associated with oral ulceration. Most oral ulcers in Behçet's syndrome resolve within 10-14 days, although replacement with new ulcers may make it feel as if they are sustained. If an ulcer is persistent, your doctor may wish to arrange a biopsy to look into the diagnosis.

Genital ulcers occur less frequently than oral ulcers. In Europe approximately 60-70 per cent of people with Behçet's Syndrome report suffering from them. They resemble oral ulcers in appearance, are usually painful and, when deep, may heal with scarring. Genital ulcers in females affect the vulva and vagina and, occasionally, the cervix. In males, ulcers occur classically on the scrotum. Because of their location, genital ulcers may become infected. Genital ulcers can be triggered by sexual intercourse and, conversely, are an obvious impediment to sex. As with oral lesions, non-healing ulcers should be biopsied.

### Skin

Skin problems are very common in Behçet's syndrome, occurring in about 80 per cent of patients. Usually people will report redness and spots (pustules) resembling acne. Steroid treatment may also cause acne which can complicate trying to decide if pustules are related to the Behçet's Syndrome or are a side effect of treatment.

Less commonly, lumps may develop on the shins that resemble bruises (erythema nodosum), or which are due to an inflamed vein under the skin.

### Pathergy

"One symptom that is partly specific for Behçet's Syndrome is the "Pathergy test". "Pathergy" is a term that describes an inappropriately large response to minor trauma. It is often experienced as prolonged or severe redness at the site of day-to-day injury, such as a needle or thorn prick or a nettle sting. The "Pathergy test" involves using a sterile hypodermic needle to make a deep prick in the forearm skin of the patient. A positive test is where a small swelling (papule), possibly with pus (pustule), is present at the site of the needle prick 24-48 hours later. If positive, the test is useful, but a significant proportion of patients with Behçet's Syndrome will have a negative test.

### Eyes



Eye inflammation may be a serious problem in some people with Behçet's Syndrome. It is thought that 50-70 per cent of people with Behçet's develop inflammation of the eye. If this is going to happen, it usually starts within 2-3

years of the diagnosis of Behçet's syndrome. It is the first symptom (or the one that leads to the diagnosis) in 10-15 per cent of cases. Eye inflammation usually comes on in unpredictable, repetitive attacks. Help should be sought quickly if any symptoms develop, as treatment may be required to avoid damage to sight.

### **Headache**

Headaches are very commonly reported in people with Behçet's syndrome. Over 80 per cent of people will report this symptom. For the vast majority, headaches are not the result of a serious problem in the brain and may improve with symptomatic treatment.

Symptoms that warrant further investigation include very severe or incapacitating pain, a change in character of the headache and particularly the presence of other symptoms (e.g. weakness, speech difficulties, visual loss). Serious brain problems occur in only a small proportion of patients.

### **Muscle and joint pains**

Pain in the muscles and joints are common in Behçet's syndrome. Occasionally joint swelling and tenderness (arthritis) occurs but this is rarely persistent. Even if arthritis occurs, it rarely leads to damage to the joints and seldom becomes a significant cause of disability.

### **Stomach pains**

Many patients with Behçet's syndrome experience abdominal pains, and symptoms similar to "irritable bowel syndrome" (bloating, diarrhoea or constipation). Camera tests such as colonoscopy / OGDs are frequently normal despite symptoms, with only about 1 in 10 symptomatic patients having visible ulceration or inflammation on these tests. Symptoms may respond to irritable bowel syndrome treatments or to treating with immunosuppressive drugs.

### **Blood clots and blood vessels**

Probably less than 1 in 10 European patients with Behçet's syndrome develop inflammation in blood vessels. The blood vessels predominantly affected are veins. Veins of any size or position may be involved, but particularly the deep veins of the legs, causing Deep Vein Thrombosis (DVT). Arteries are less commonly affected. Aneurysms (expansion of arteries) can occasionally occur.

There is no good evidence that Behçet's syndrome predisposes to ordinary heart disease.

### **Tiredness**

Most patients with Behçet's syndrome report feeling fatigued and this may well impact on quality of life. The reasons for fatigue are often complex, often including inflammation, loss of physical fitness, pain, low mood and/or poor sleep. In addition to treatments to suppress the inflammation, attempts to increase fitness (e.g. swimming or pilates), address sleep disturbances, pain and mood may improve this symptom.

### **Making a diagnosis**

No diagnostic laboratory or imaging test currently exists for Behçet's syndrome. A doctor will probably wish to undertake blood tests to help rule out other types of illness and to record the amount of inflammation. However, "inflammatory marker" blood tests (eg ESR, CRP) may be normal or only modestly raised despite ongoing clinical symptoms.

One blood test that may be helpful is "HLA-B51 expression". HLA B51 is a protein that is found on the surface of blood cells. People differ in the exact HLA proteins on the surface of their cells, and 'HLA B51' is the one found quite frequently in people with Behçet's Syndrome. It is important to realise that many people with Behçet's syndrome will not have this protein, and, equally well, most people who have this protein will never develop Behçet's Syndrome.

The diagnosis therefore has to be decided on clinical grounds (i.e. on the basis of the opinion that your doctor forms after talking to you, examining you, and after taking blood tests to rule out other illnesses).

### **Clinical classification criteria**

Clinical classification criteria were developed by a group of doctors in 1990 for standardising patients for research studies. It is possible to still have Behçet's syndrome without fully fulfilling these research criteria.

### **The Classification criteria published by the International Study Group in 1990**

1. Oral ulcerations occurring at least three times a year
2. Plus two of the following in the absence of other systemic diseases:
  - recurrent genital ulcers
  - specific eye lesions observed by an ophthalmologist
  - skin lesions consistent with Behçet's syndrome

### **Treatment**

#### **Ulcers**

The aim of therapy is to reduce the frequency and severity of ulcers to an acceptable level. Topical measures should be tried first when possible, particularly in mild disease. Focusing on oral hygiene by using chlorhexidine mouth washes to reduce bacterial growth may reduce the frequency of mouth ulcers and subsequent infection. Topical corticosteroids are often helpful, as suggested by their widespread use rather than by robust clinical trials. These agents can be applied in many ways, including a beclomethasone inhaler (designed for asthma) to spray onto incipient lesions as they first become symptomatic, triamcinolone acetonide in adhesive bases or hydrocortisone lozenges.

Local steroid preparations may also be applied to genital ulcers, using an inhaler spray or cotton tips. Sucralfate can be used as an oral rinse or directly applied to genital ulcers.

Colchicine is usually the first-line tablet therapy for orogenital ulceration when topical measures are inadequate. Many patients will require further escalation in therapy and azathioprine is the usual first choice. If this therapy fails, an alternative can be chosen from the list below with due consideration of toxicity and cost.

#### **Drugs and Side effects**

For information on the main drugs prescribed for Behçet's Syndrome see:

- Colchicine
- Azathioprine
- Methotrexate
- Steroids
- Cyclophosphamide (for severe illness)
- Ciclosporin (no longer in common use)

Also used to treat Behçet's - Interferon therapy. Thalidomide (no longer in common use)

For information on other drugs used in the treatment of vasculitis see: Glossary of drugs and side effects - [www.vasculitis.org.uk/about-vasculitis/glossary-of-drugs](http://www.vasculitis.org.uk/about-vasculitis/glossary-of-drugs)

#### **General considerations**

Lifestyle modifications to reduce stress should be considered as anecdotal experience suggests that stress exacerbates Behçet's syndrome. It is often worthwhile checking for and correcting vitamin D deficiency, as vitamin D has many immune-modulating effects and is often at surprisingly low levels.

#### **Surgery and Behçet's Syndrome - what should I know?**

Given the large inflammatory response that people with Behçet's syndrome mount to trauma, surgery should not be undertaken lightly. A course of steroids, such as prednisolone, starting 7-10 days preoperatively and reducing rapidly after the procedure may be an option that you may wish to discuss with your specialist and surgeon, as they may help to prevent inflammatory complications developing in the perioperative period. However, the risk benefit ratio needs to be considered, given that steroids also may increase risk of infection. Arterial catheterisation is best avoided when possible, for fear of aneurysm development at the arterial entry site. People often report that visits to the dentist or oral hygienist result in an outbreak of oral ulcers, which can often be avoided with a short course of steroids prior to the visit.

#### **Pregnancy and Behçet's syndrome**

There is no evidence that people with Behçet's syndrome have an increased risk of fertility problems, or complications during pregnancy, although women with vascular, neurological or eye involvement need to have a serious discussion with their doctors before proceeding. It is very important to discuss medications with your doctor

as many of the medications that patients are treated with may potentially harm a developing baby if not stopped well in advance of getting pregnant.

### **Prognosis**

For the vast majority of people with Behçet's syndrome, a normal life span is expected. For those with severe eye inflammation, progressive loss of vision was previously a major complication. However, with improving treatments, visual loss is becoming more uncommon. Those with purely 'skin/ulcers/joint' disease have a very good prognosis.

### **Key Points**

- Behçet's syndrome is a multisystem condition, the hallmark features being mouth and genital ulcers, although many areas may be affected, including inflammation in the eyes
- Behçet's syndrome commonly affects the skin, stomach and musculoskeletal systems
- No diagnostic test currently exists for Behçet's syndrome
- Treatment is based on immunosuppression with agents including steroids, azathioprine, and anti-TNF biologic agents

### **Acknowledgements**

Vasculitis UK is indebted to Dr Nicola Ambrose, MB BCh, MRCPI, PhD for producing this web page on Behçet's syndrome. Dr Ambrose is a Rheumatologist working at University College London Hospital, having received her training in Behçet's syndrome working with Prof Dorian Haskard.

### **Related Vasculitis Articles**

- Fertility and Vasculitis - Dr David Jayne - [www.vasculitis.org.uk/living-with-vasculitis/fertility](http://www.vasculitis.org.uk/living-with-vasculitis/fertility)

### **Further reading**

- Behçet's Disease (Adamantiades-Behçet's Disease) - Fumio Kaneko, Ari Togashi, Sanae Saito, et al - [www.ncbi.nlm.nih.gov/pmc/articles/PMC2967828/pdf/CDI2011-681956.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2967828/pdf/CDI2011-681956.pdf)
- Alemtuzumab (CAMPATH-1H) as Remission Induction Therapy in Behçet's Disease - [www.blackwellpublishing.com/acrmeeting/abstract.asp?MeetingID=774&id=89925](http://www.blackwellpublishing.com/acrmeeting/abstract.asp?MeetingID=774&id=89925)
- Vasculitis Paediatric Guidelines 2012 - Dr Helen Foster & Dr Paul Brogan, Oxford University Press - <http://www.vasculitis.org.uk/professionals/paediatric-vasculitis-guidelines>

### **Behçet's - Centres of Excellence**

In February 2012 three UK Behçet's Centres of Excellence, for the diagnosis and treatment of Behçet's Disease, were announced. These are to be at Bart's and the London Hospital, Birmingham City Hospital and at Aintree University Hospital in Liverpool. The centres will be lead by Prof Farida Fortune (London), Dr Deva Situnayake (Birmingham) and Prof Robert Moots (Liverpool).

**UK Behçet's Society** [www.behcets.org.uk](http://www.behcets.org.uk)

**American Behçet's Disease Association** [www.behcets.com](http://www.behcets.com)

# Buerger's Disease

## What is Buerger's Disease?

Buerger's (also known as Thromboangiitis obliterans) is a disease of the small and medium arteries and veins that restricts blood flow to the hands and feet. Clots (thrombus) develop inside the blood vessels. This in turn leads to skin ulcers and gangrene in the fingers and toes and numbness and tingling if the nerves are affected. It is not uncommon for toes, fingers and limbs to need to be amputated if the gangrene progresses. It is not clear whether this is a true vasculitis.

## Who are affected?

The disease is seen almost exclusively in smokers, mainly in young men aged 20-40 years. Recently, however, a higher percentage of women and patients over 50 have been diagnosed.

## What are the symptoms?

The initial symptoms include claudication (pain induced by insufficient blood flow during exercise) in the feet and or the hands. The pain usually begins in the extremities but may radiate to other parts of the body. Patients may experience numbness and tingling in the limbs and also Raynaud's Phenomenon. Raynaud's is a condition where the extremities of the hands and feet turn white when exposed to cold. A common sign in Buerger's is skin ulceration and gangrene of the fingers and toes.

## What is the aetiology (cause)?

Use of tobacco, particularly cigarette smoking, is the overwhelming factor predisposing to a diagnosis of Buerger's Disease. The majority of patients are heavy smokers, although some cases have been reported in moderate smokers and even in patients using chewing tobacco or other forms of tobacco that are not smoked. It is thought that this disease is triggered by some constituent of tobacco.

## Diagnosis

Angiograms of the limbs can be helpful in making a diagnosis of the disease. Biopsies are not usually recommended as a diagnostic tool for Buerger's Disease because of the possibility that the biopsy area may not heal well. Typically blood tests are normal and do not show any evidence of inflammation.

## Treatment

It is essential that people affected with Buerger's Disease stop smoking completely and immediately. Continued smoking or use of other forms of tobacco, even small amounts, increases the risk of losing fingers, toes or limbs due to necessary amputation.

Although there is evidence of inflammation inside the vessels and within the blood clots this differs from other types of vasculitis where the inflammation is in the blood vessel wall. Anti-inflammatory and immunosuppressant treatments have not been shown to be effective in Buerger's Disease. Other treatments such as iloprost or prostacyclin that help to open up the blood vessels may be helpful.

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

## Prognosis

Good with correct treatment and complete cessation of smoking and tobacco use.

## Key points

- Buerger's disease is associated with the use of tobacco
- The most important treatment is stopping smoking.

## Further reading

Buerger's disease/Thrombotic obliterans - Gregory Piazza, Mark A. Creager  
<http://circ.ahajournals.org/content/121/16/1858.full.pdf+html>

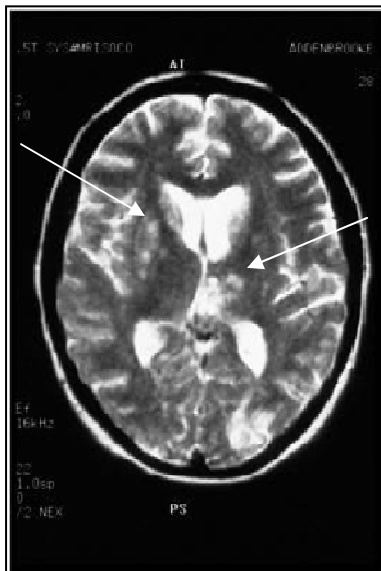
## Central Nervous System Vasculitis

### What is Central Nervous System Vasculitis?

Many different types of vasculitis can affect the blood vessels in the brain (called Central Nervous System Vasculitis (CNS)) including the ANCA associated vasculitides, Takayasu arteritis and Giant Cell Arteritis.

### Primary Angiitis of the Central Nervous System

When inflammation of the blood vessels of the brain is found in the absence of another cause, including other types of vasculitis or inflammatory diseases, infection or cancer it is usually called Primary Angiitis of the Central Nervous System (PACNS). It is an inflammatory disease affecting the arteries of the brain causing a wide variety of symptoms.



### Primary cerebral vasculitis confirmed by brain biopsy

Arrows indicate abnormal areas

Image courtesy of Dr David Jayne, Addenbrooke's Cambridge, UK

### Who are affected?

PACNS affects males more than females. The usual age of onset is the fourth or fifth decade of life but children can be affected as can much older patients.

### What is the aetiology (cause)?

The cause of PACNS is not yet known.

### What are the symptoms?

The common symptoms of CNS are: confusion, headache and personality change. Other symptoms noted are: seizures, bleeding in the head, coma and vision loss. Occasionally patients can develop symptoms similar to stroke. Usually these symptoms appear over several months but can occur quickly.

### Making a diagnosis

Because of the slow onset of symptoms it can take several months for the diagnosis of PACNS to be made. Many other diseases including infections and cancers can mimic PACNS and these must be excluded. The most definite way to make the diagnosis is a brain biopsy showing evidence of inflamed blood vessels. Unfortunately this test can often be misleadingly negative. MRI and CT scans are usually abnormal but the findings are not specific for PACNS. Blood tests are often normal but may show evidence of inflammation.

### Treatment

As with other types of serious vasculitis high doses of steroids and cyclophosphamide are often used to bring the disease under control (establish remission). Lower dose immunosuppression is then used to maintain remission in the long term.

### Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

### Prognosis

Relapse occurs in about a quarter of cases. With correct treatment the prognosis is much better than in the past.



### **Key points**

- PACNS is very rare
- Symptoms are non-specific and may develop over a long period of time
- Treatment depends on disease severity.

### **Further reading**

**Diagnosis and treatment of cerebral vasculitis** - Peter Berlit

[www.ncbi.nlm.nih.gov/pmc/articles/PMC3002614/pdf/10.1177\\_1756285609347123.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3002614/pdf/10.1177_1756285609347123.pdf)

**Primary Angiitis of the Central Nervous System** - Julius Birnbaum, MD; David B. Hellmann, MD

<http://archneur.jamanetwork.com/searchresults.aspx?q=primary%20angiitis%20of%20the%20central%20nervous%20system&t=&p=1&s=1&c=0>

## Cogan's Syndrome

### What is Cogan's Syndrome?

Cogan's Syndrome is a rare inflammatory disease characterised by inflammation of the inner ears and eyes. It can lead to vision difficulties, hearing loss and dizziness. Commonly there is also inflammation in other organs as well, particularly the heart and large blood vessels, nervous system and bowels,

### Who are affected?

Although any age can be affected the syndrome is commonest in young adults (20's and 30's). It affects males and females equally.

### What is the aetiology (cause)?

The cause of Cogan's Syndrome is not yet known.

### What are the symptoms?

The most common symptoms include red, painful, light-sensitive eyes or blurred vision; hearing loss (which may become profound and permanent); vertigo (dizziness); poor balance; nausea and vomiting; fever, fatigue and weight loss.

### Making a diagnosis

There are no specific diagnostic tests. The diagnosis is made on clinical examination and history where a combination of problems in the eyes and inner ears are described. Other infections/diseases, including Wegener's Granulomatosis and Rheumatoid Arthritis need to be excluded.

### Treatment

There are no clinical trials of treatment in Cogan's Syndrome. Most people with Cogan's Syndrome will need treatment with moderately high doses of prednisolone or other types of steroids. A few patients with very mild eye disease may be treated with anti-inflammatory drugs including steroids and nonsteroidal anti-inflammatory drugs (NSAIDs) which are applied to the eye. Many patients will also require additional treatment with other immunosuppressive drugs including Methotrexate, Cyclosporin, Azathioprine, Tacrolimus or Cyclophosphamide.

### Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

### Prognosis

The course of the disease varies significantly from patient to patient. In some patients there is an initial flare, which may last several weeks to months. Following this there may be a slowly progressive course in some patients while others have a course of complete remission with intermittent episodes of disease activity. Fortunately blindness occurs in less than five per cent of patients. Deafness is a frequent and debilitating outcome occurring in up to 54 per cent of patients.

### Key Points

- Cogan's Syndrome commonly affects the eyes and ears causing vision, hearing and balance problems
- Treatment will depend on the disease severity.

### Further reading

#### A shifty diagnosis: Cogan's syndrome. A case report and review of the literature -

G Migliori, E Battisti, M Pari, N Vitelli, C Cingolani

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2808685/pdf/0392-100X.29.108.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2808685/pdf/0392-100X.29.108.pdf)

# Cryoglobulinemia and Cryoglobulinaemic Vasculitis

## What is Cryoglobulinemia?

Cryoglobulinemia means "cold antibody in the blood" and is the presence of abnormal antibodies that are soluble in the blood at body temperature but which precipitate out of the blood at lower temperatures in the laboratory. These antibodies are often present in patients with a wide variety of pre-existing diseases such as hepatitis C virus infection, autoimmune diseases such as rheumatoid arthritis or Sjögren's Syndrome or cancers such as lymphoma or multiple myeloma. In the very rare cases where an underlying disease is not identified the presence of cryoglobulin antibodies in the blood is called "essential cryoglobulinaemia". Often the antibodies do not cause any problems and then no treatment may be needed. In some patients they cause a vasculitis of the small and medium blood vessels called Cryoglobulinaemic Vasculitis. This condition may need treatment.

## What are the symptoms?

Cryoglobulinaemic Vasculitis symptoms include rash on the lower limbs, joint pain or arthritis, nerve damage, abdominal pain or kidney failure.

## What is the aetiology (cause)?

The commonest underlying causes of cryoglobulinaemia are hepatitis C infection, lymphoma and myeloma. The cause of essential cryoglobulinaemia is not known and can only be diagnosed once all the possible underlying causes have been excluded.

## Making a diagnosis

Diagnosis is made by recognising the clinical symptoms and signs and performing the specific laboratory test for cryoglobulins. A biopsy of the affected tissue or organ may be necessary to confirm the diagnosis.

## Treatment

The most important treatment is of the underlying disease and will depend on the correct diagnosis. Patients with severe forms of vasculitis may need additional treatment with steroids, immunosuppressant drugs or plasma exchange to control the vasculitis.

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

## Prognosis

Prognosis is determined by the underlying disease as well as other factors including age, and the severity of damage to any organs, especially the kidneys.

## Key Points

- Cryoglobulins and cryoglobulinaemic vasculitis are often caused by underlying diseases including infections and cancers
- Treatment depends on the underlying disease and the severity of the vasculitis.

## Further reading

Mixed Cryoglobulinemia - Clodoveo Ferri

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2569912/pdf/1750-1172-3-25.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2569912/pdf/1750-1172-3-25.pdf)

Cryoglobulinaemic vasculitis: classification and clinical and therapeutic aspects - Gerald S Braun, Sophia Horster et al [www.ncbi.nlm.nih.gov/pmc/articles/PMC2805946/pdf/87.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2805946/pdf/87.pdf)

## Hypersensitivity Vasculitis

### What is Hypersensitivity Vasculitis?

Hypersensitivity vasculitis (HV) is a term often used to describe many different conditions. Most commonly the term is applied to a vasculitic skin rash caused by sensitivity to a drug although it may be seen in association with several other conditions. Most commonly only the skin is affected although the bowels, kidneys and joints may also be affected.

### Who are affected?

HV usually affects adults, although children have been found to have the condition.

### What are the symptoms?

The commonest symptoms are those related to the skin. Several different types of rash are recognised as HV. The commonest types of rash are small or large raised purpled spots and flat red areas. Symptoms in the skin include burning, pain or itching or the rash may not have any symptoms. Joint involvement may cause painful joints. Bowel involvement may cause a variety of symptoms including abdominal pain and diarrhoea. Kidney involvement may lead to blood in the urine or kidney failure.

### What is the aetiology (cause)?

No cause for HV is found in between one third and one half of patients. HV can be caused by a specific drug or by infection. The most common drugs include: antibiotics (particularly penicillins), non-steroidal anti-inflammatory drugs (NSAIDs) and diuretics. Infections which may be associated with HV include: hepatitis B or C virus, chronic infection with bacteria or HIV. HV type rashes and symptoms may also be seen as part of other inflammatory disease including other types of vasculitis, Systemic Lupus Erythematosus (SLE), Rheumatoid Arthritis, Sjögren's Syndrome, inflammatory bowel disease and very rarely some types of cancer.

### Making a diagnosis

There are no specific tests for HV. Blood tests may show evidence of inflammation. Tests are usually performed to determine if the patient has one of the diseases or infections listed in the section above. If a new drug has been recently started it may suggest that this has caused the HV. Sometimes a skin biopsy is necessary to prove that there is a vasculitis

### Treatment

If drugs are implicated as the cause then the drugs should be stopped. Where infection is the cause treatment of the infection is required. If another inflammatory disease is the cause this should be treated appropriately. Patients with skin rash and joint disease may respond to dapsone or colchicine. Some patients, particularly those with severe kidney or bowel involvement, may require treatment with steroids and other immunosuppressant drugs.

### Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

### Prognosis

For patients with HV caused by an identified drug or treatable infection the prognosis may be good and the disease may not recur. For patients with other underlying inflammatory diseases, chronic infections, cancers and for where no cause is found the prognosis may depend on the underlying disease and the condition may recur.

### Key points

- HV is usually caused by medications or other underlying diseases
- Treatment and prognosis depend on identifying the causative factors where possible.

# Rheumatoid Vasculitis

## What is Rheumatoid Vasculitis?

Rheumatoid Vasculitis (RV) is a serious complication of rheumatoid arthritis where there is inflammation of the blood vessels. It commonly affects the skin causing rash and ulcers and the nerves causing loss of sensation although any part of the body may be involved.

## Who are affected?

RV is reported to be commoner in men with rheumatoid arthritis (1 in 9 men in one study) than in women (1 in 38 women in one study) but appears to be becoming much rarer over the last few decades. Patients with rheumatoid factor antibodies and rheumatoid nodules are the patients most likely to develop RV. Patients who develop Felty's Syndrome (a combination of rheumatoid arthritis, large spleen and low neutrophil count in the blood) are also more likely to develop RV. There is also an association with some genetic inheritances and smoking.

## What are the symptoms?

The symptoms will depend upon the part of the body affected. Skin rashes and ulcers are a common problem. Involvement of the nerves usually causes numbness or tingling and "pins and needles" symptoms, muscle weakness may also develop. Inflammation may develop around the outside of the heart (pericarditis) which can cause chest pain. Inflammation in the eyes may cause pain, redness and impaired vision. Rarely the kidneys may be involved causing blood and protein in the urine or kidney failure. Non-specific features may include fatigue, fevers and weight loss.

## What is the aetiology (cause)?

The cause of RV is not yet known. In RV it has been noted that there are more likely to be particular types of antibodies in the blood and they will be at a higher level than in rheumatoid arthritis patients without RV. A link with smoking has been noted and some researchers suggest that smoking leads to damage to the lining of the blood vessels that pre-disposes to developing RV.

## Making a diagnosis

There is no specific test for diagnosing RV. Blood tests may show evidence of inflammation and high levels of some antibodies but these are not specific for RV. A biopsy of an affected tissue or organ (skin, muscle, nerve, kidney etc) may confirm vasculitis but again may not be specific for RV and other types of vasculitis should be excluded. Neurophysiology tests (electrical tests of nerves and muscles) may show evidence of inflammation and sometimes be used to guide a biopsy. Angiography (x-ray or other imaging of the blood vessels) may show non-specific evidence of vasculitis.

## Treatment

Treatment depends on the severity of the vasculitis and the organs involved. Mild RV affecting only skin or peripheral nerves may be treated with steroids and immunosuppressants such as methotrexate or azathioprine. Severe RV may need treatment with high dose steroids and cyclophosphamide. Newer drugs such as anti-TNF antibodies and rituximab have also been successfully used in RV in combination with other drugs.

For RV involving the fingertips, skin around the fingernails or in the case of a rash the treatment is pain control, antibiotic cream and local protection. **Stopping smoking is essential.**

## Side effects

For the side effects of the drugs used see "Glossary of drugs and side effects".

## Prognosis

The prognosis in RV depends in part on the severity of the damage to the organs involved. RV may also increase the risk of developing cardiovascular disease. Some previous studies have suggested that the development of RV in a patient with rheumatoid arthritis may have a significantly worse prognosis than rheumatoid arthritis patients without RV.

## Key points

- RV is part of rheumatoid arthritis in some patients
- Treatment and prognosis depend on the disease severity
- Stopping smoking.

## Further reading

### **Rheumatoid Vasculitis: Vanishing Menace or Target for New Treatments? -**

C. M. Bartels, A. J. Bridges

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2950222/?tool=pubmed](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2950222/?tool=pubmed)

## Videos

### **Rheumatoid Vasculitis** Professor David G I Scott

<http://www.excellence-in-rheumatology.org/content/rheumatoid-vasculitisdavid-giscott?mid=53>

### **What is the relationship between vasculitis and rheumatoid arthritis (RA)? -**

**Professor David GI Scott**

[www.excellence-in-rheumatology-institute.org/content/85/what-relationship-between-vasculitis-and-rheumatoid-arthritis-ra-prof-david-gi-scot?mid=50](http://www.excellence-in-rheumatology-institute.org/content/85/what-relationship-between-vasculitis-and-rheumatoid-arthritis-ra-prof-david-gi-scot?mid=50)

## Urticarial Vasculitis

### What is Urticarial Vasculitis?

Urticarial Vasculitis is a form of cutaneous vasculitis characterised by inflammation of the small blood vessels. Urticarial Vasculitis can be classified into three subtypes. All are defined by a measure of the "complement" levels in the blood. The complement system is a set of proteins that contribute to and amplify immune responses. They play a role in some, but not all, autoimmune disorders including some forms of Urticarial Vasculitis.

### Normocomplementemic Urticarial Vasculitis (NUV)

NUV is diagnosed where a patient has the main symptoms of Urticarial Vasculitis combined with normal levels of C1q complements. NUV is generally the least severe form of Urticarial Vasculitis. It is less likely to be associated with any other symptoms.

### Hypocomplementemic Urticarial Vasculitis (HUV)

HUV is diagnosed where the patient has the main symptoms of Urticarial Vasculitis combined with lower than normal levels of C1q complements and raised levels of anti-C1q antibodies (antibodies that attack the C1q complements). HUV is a more severe form of Urticarial Vasculitis and is likely to include symptoms such as purple or dark red spots or rash on the skin (a typical vasculitic rash); arthritic joint pain; breathing difficulties such as asthma, and stomach pains. Some regard HUV as a form of SLE (Lupus), although HUV patients usually test Anti-Nuclear Antibody (ANA) negative instead of the normal positive result for Lupus patients.

### Hypocomplementemic Urticarial Vasculitis Syndrome (HUVS)

HUVS is a very rare and severe form of Urticarial Vasculitis. HUVS patients have more extensive complement abnormalities (low circulating 3rd and 4th complement components). As well as the symptoms of HUV, patients will suffer from systemic problems such as: episcleritis or uveitis (bloodshot or inflamed eyes) (found in 30 per cent of patients); mild glomerulonephritis (kidney disease); pleuritis (inflammation of the membrane surrounding the lungs); angioedema (swelling of the tissues under the skin) (found in 50 per cent of patients); Chronic Obstructive Pulmonary Disease (COPD) (breathing difficulties) (found in 50 per cent of patients), and cardiac involvement such as myocardial infarction (heart attack)

### Who are affected?

NUV is most common between the ages of 30-40 and is found in women more than men. HUV and HUVS are almost exclusively found in women.

### What is the aetiology (cause)?

The cause of Urticarial Vasculitis is not known in 50 per cent of cases. However it can be associated with or triggered by autoimmune/connective tissue diseases like Systemic Lupus Erythematosus (SLE or simply Lupus) Rheumatoid Arthritis, or occur in the context of a systemic vasculitis syndrome, such as Churg Strauss syndrome (Eosinophilic Granulomatosis with Polyangiitis); infections or viruses such as hepatitis; a reaction to certain drugs such as ACE inhibitors; certain types of diuretics; penicillin and Non-Steroidal Anti-Inflammatory Drugs (NSAIDs); cancer; and Glandular problems, such as Graves' Disease (overactive thyroid).

### What are the symptoms?

The main symptom is a recurring urticarial ("stinging nettle") rash that lasts for longer than 24 hours and "burns" rather than "itches", leaving marks behind on the skin when it clears.



◀ Stinging nettle rash

## Diagnosis

As in other types of vasculitis, diagnosis will depend on the doctor recognising the pattern of symptoms and examination findings. Blood is commonly taken to test for raised levels of "C-reactive protein" (CRP) and "Erythrocyte Sedimentation Rate" (ESR) which indicate inflammation in the body. Skin and kidney biopsies may also be taken to confirm the diagnosis.

Testing for C1q, anti-C1q, C3 and C4 complement components is essential to determine the type of Urticarial Vasculitis.

Tests may also be done for Anti-Nuclear Antibody (ANA) levels (which are positive in 30 to 50 per cent of patients), and Anti ds-DNA levels may also be positive.

## Treatment

Milder cases of Urticarial Vasculitis may simply be treated with antihistamines and NSAIDs such as Ibuprofen. Corticosteroids such as Prednisolone might be used for more persistent cases. However for the more severe forms of Urticarial Vasculitis, steroids such as Prednisolone are usually prescribed to reduce inflammation as well as immunosuppressants such as Azathioprine, Cyclophosphamide or Mycophenolate Mofetil (CellCept).

In cases where a patient is unresponsive to treatment, intravenous immunoglobulin and anti-cytokine monoclonal antibodies or rituximab may have a role.

If the disease is very severe large doses of Methylprednisolone or Plasmapheresis (plasma exchange) may also be given. When the disease becomes quiet less toxic drugs are used to keep control and these include: Azathioprine, Methotrexate and Mycophenolate Mofetil usually in combination with low dose prednisolone.

## Prognosis

The overall prognosis in Urticarial Vasculitis depends on the severity of the disease and the amount of damage that has been done to organs, especially the lungs. The main risk to patients appears to be Chronic Obstructive Pulmonary Disease (COPD).

Smoking is a major risk factor for fatal lung disease in HUVS and smokers should seek help to give up as soon as possible.

## Key Points

- The systemic HUVS type of Urticarial Vasculitis is a very rare form of the disease
- The main symptom is a recurring urticarial ("stinging nettle") rash
- The levels of C1q complements in the blood are a significant indicator for diagnosis and treatment
- Treatment depends upon the severity of the disease

## Further reading

Hypocomplementemic Urticarial Vasculitis Syndrome. Clinical and serological findings in 18 patients - Wisnieski JJ, Baer AN et al <http://www.ncbi.nlm.nih.gov/pubmed/7837968/>

Vasculitis: Approach to diagnosis and therapy - Aparna Palit, Arun C Inamadar <http://www.bioline.org.br/pdf?dv06117>

Hypocomplementemic Urticarial Vasculitis Syndrome - An Interdisciplinary Challenge - Wolfgang Grotz, Hideo A Baba et al <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2795336/>



## Anti-GBM (Goodpasture's Disease)

The Trust is indebted to Professor Charles Pusey, Professor of Medicine and Head of Section, Renal Section, Imperial College, London, for providing the following information on Anti-GBM (Goodpasture's Disease)

### What is anti-GBM disease?

Anti-glomerular basement membrane (GBM) disease, also known as Goodpasture's disease, is a rare condition that causes inflammation of the small blood vessels in the kidneys and lungs.

### Who are affected?

This disease tends to affect two age groups - young people aged 20 to 30, and older people in their 60s and 70s. Men are affected slightly more often than women.

### What is the aetiology (cause)?

The disease occurs when the body's immune system attacks the lining (known as the glomerular basement membrane, GBM) of the small blood vessels in the kidneys and lungs. It is not currently known why this occurs, although people with a particular "tissue type" seem to be at greater risk. Lung involvement is more common in cigarette smokers, and it is thought smoking may also be a trigger for developing the disease.

### What are the symptoms?

Anti-GBM disease usually develops suddenly, over a period of a few weeks. The kidneys, the lungs, or both may be affected. About 50 per cent of patients have lung disease in addition to kidney disease, and isolated lung disease is very rarer. Kidney involvement may not cause symptoms at first, although blood and protein may be detected in the urine. As the disease progresses, patients may develop signs of kidney failure such as tiredness, poor appetite, decreased urine production, breathlessness and leg swelling. When the lungs are involved, patients may have severe breathless, dry cough, or coughing up blood.

### Making a diagnosis

The diagnosis of anti-GBM disease usually relies on a blood test to identify anti-GBM antibodies. Often a kidney biopsy (taking a small sample of kidney tissue with a needle) is required to demonstrate that the kidneys are involved. This may also confirm the diagnosis, by showing deposition of anti-GBM antibodies in the kidney. Other tests (such as X-rays or CT scans) are often used to show if the lungs are involved.

### Treatment

Treatment requires a process called plasma exchange, which involves the use of a machine to remove anti-GBM antibodies from the bloodstream. This is often done daily for 2 weeks. In addition, immunosuppressive drugs such as steroids and cyclophosphamide, are used to suppress inflammation and stop further antibody production. Treatment usually continues for 6 months after the diagnosis is made.

Details about plasma exchange can be found under Glossary of Procedures: [www.vasculitis.org.uk/about-vasculitis/glossary-of-procedures](http://www.vasculitis.org.uk/about-vasculitis/glossary-of-procedures)

The treatment of vasculitis can be found under Glossary of Drugs - [www.vasculitis.org.uk/about-vasculitis/glossary-of-drugs](http://www.vasculitis.org.uk/about-vasculitis/glossary-of-drugs)

### Prognosis

When treatment is started promptly, before the need for dialysis, the majority of patients recover to have normal kidney and lung function. However, patients already on dialysis are unlikely to recover kidney function. Unlike some other forms of vasculitis, it is very rare for anti-GBM disease to relapse, and thus it does not usually require long-term treatment. Smoking is thought to be a possible trigger for causing relapses, and should be avoided. In patients remaining on dialysis, kidney transplantation is possible once anti-GBM antibodies have become undetectable.

### Key points

- Anti-GBM disease is a rare form of vasculitis
- It causes damage to small blood vessels in the kidneys and lungs
- Diagnosis is confirmed by detecting anti-GBM antibodies in blood or deposited in the kidney

- Treatment with plasma exchange (to remove anti-GBM antibodies) together with prednisolone and cyclophosphamide is usually successful

#### **Further reading**

- The Kingston Whig - Rare condition will cost ailing teenager her kidneys - [http://www.thewhig.com/2012/12/10/rare-condition-will-cost-ailing-teen-her-kidneys?utm\\_source=addThis&utm\\_medium=addthis\\_button\\_facebook&utm\\_campaign=Rare+condition+will+cost+ailing+teen+her+kidneys+|+Local+|+News+|+The+Kingston+Whig-Standard#.UR6wT91M7w.facebook](http://www.thewhig.com/2012/12/10/rare-condition-will-cost-ailing-teen-her-kidneys?utm_source=addThis&utm_medium=addthis_button_facebook&utm_campaign=Rare+condition+will+cost+ailing+teen+her+kidneys+|+Local+|+News+|+The+Kingston+Whig-Standard#.UR6wT91M7w.facebook)

**Vasculitis images** - all courtesy of Vasculitis UK Members and vasculitis patients



GPA patient undergoing peritoneal exchange "on the move"



Vasculitis rash



Necrosis of the toes in a GPA patient



Mouth ulcer in Behçet's



GPA rash in a patient not responding to Cyclophosphamide



Back rash in unspecified vasculitis  
Image courtesy of David M Bryan



Tracheostomy tube



Tracheostomy tube neck band customised by patient



Leg rash in Takayasu Arteritis

**Vasculitis images continued:**



Infected biopsy site in HSP



Skin involvement in a patient with EGPA



3 illustrations of skin rash in a patient with unspecified pANCA vasculitis



Behçet's rash



Rash in LCV  
(Leukocytoclastic vasculitis)

## Glossary of drugs and side effects

### Steroids and Immunosuppressants

Both Prednisolone (steroids) and Immunosuppressants such as Cyclophosphamide, Azathioprine, Methotrexate and Mycophenolate Mofetil reduce inflammation by suppressing the activity of the white blood cells. However, by doing this these white blood cells are less able to fight infection. The immunosuppressants can also suppress the production of cells from the bone marrow.



If necessary blood tests will be performed to monitor closely the white blood count and haemoglobin (to detect anaemia). If a patient has signs of a fever or infection it is imperative that they seek medical attention.

**Not all the side effects noted with these drugs are mentioned below. Patients are advised to read the patient leaflet supplied with the drugs, and speak to their physician/pharmacist if they are unsure about anything contained in the leaflet.**

### List of drugs and side effects used to treat vasculitis

The following is a list of many of the drugs used to treat and maintain remission in the various vasculitides. Some of the drugs mentioned are prophylactic (prescribed to prevent disease). A number of the drugs are general to many of the vasculitides whilst others are specific to individual vasculitic diseases.

The list is not exhaustive and new drugs are constantly being developed. The lists of side effects is not exhaustive and patients are encouraged to read the information sheets supplied with prescription medication and to discuss any concerns with their medical team.

### Immune suppressant drugs

#### Cyclophosphamide

Cyclophosphamide is commonly used to treat severe forms of vasculitis and is also widely known as an anti-cancer chemotherapy drug. It is usually given as either a daily tablet or intermittent injection every few weeks. Cyclophosphamide has the potential to cause serious short term side effects including bladder irritation (cystitis) and bone marrow suppression (low white cell count) leading to infection. Mesna (a uroprotectant) is often given to patients receiving injections of cyclophosphamide to reduce the risk of bladder problems. Patients may also be advised to drink a lot of water to flush out the bladder but this should be discussed with the medical team. Other short term effects include nausea and vomiting and hair loss. Over a longer period of time cyclophosphamide can cause infertility in both men and women. Men who may still want to have children should consider sperm banking. Long term use of cyclophosphamide is considered undesirable and has been linked to an increased risk of some types of cancer, eg cancer of the bladder.

**Monoclonal antibodies** - these include Adalimumab (Humira), Infliximab and Rituximab :

#### Adalimumab (Humira)

An artificial manufactured antibody that can be injected to reduce the effect of tumour necrosis factor alpha (TNF). TNF is an important protein in the body that can cause inflammation and is important in fighting infection. Immediate side effects are unusual but can include: mouth ulceration, diarrhoea, cough, dizziness, fatigue, paraesthesia (tingling of the skin), musculoskeletal pain, rash and pruritus (itching). A major side effect can be a reduced ability to fight some types of infection and possibly to control cancer cells in the body.

#### Infliximab

An artificial manufactured antibody that can be injected to reduce the effect of tumour necrosis factor alpha (TNF). TNF is an important protein in the body that can cause inflammation and is important in fighting infection. Immediate side effects are unusual but can include: diarrhoea, flushing, chest pain, dyspnoea (shortness of breath), dizziness and fatigue. A major side effect can be a reduced ability to fight some types of infection and possibly to control cancer cells in the body.

### **Rituximab**

A manufactured artificial antibody which removes B-cells (a type of white cell). It is now increasingly being used in patients as an alternative to cyclophosphamide and other immunosuppressants in the types of vasculitis where B cells and the antibodies they produce are thought to be important. Side effects are generally mild and are infusion related – occurring during or up to 2 hours after infusion. Side effects include: rashes and gastro intestinal (GI) upset.

### **Steroids** (Prednisolone, Methylprednisolone, Cortisone, Prednisone, Dexamethasone)

Steroids are naturally occurring hormones produced in the body by the adrenal glands and are essential for normal health. Artificial steroids are commonly used to treat vasculitis and other inflammatory diseases and are very effective at reducing inflammation. They are often used initially at high doses to control the disease and then the dose will be reduced as quickly as is possible to reduce the side effects. Unfortunately side effects are very common with steroid medication. Common serious side effects include increased risk of infection, diabetes and osteoporosis. Other common side effects include weight gain, disturbed sleep, altered mood (including rarely depression and very rarely psychosis), muscle weakness, dyspepsia (indigestion) and stomach ulcers, increased hair growth, fluid retention, increased blood pressure and thin skin. When artificial steroids are being taken the adrenal glands may stop producing the body's own steroids so it is essential that the patient does not stop taking steroids suddenly and the rate of reduction must be closely monitored by the physician. Stopping steroids too quickly can result in steroid deficiency if the adrenal glands are unable to respond quickly and this can cause significant problems. It is essential that patients on high dose steroids are monitored for the development of diabetes. If appropriate, patients may be prescribed bisphosphonates, calcium or vitamin D to protect their bones. Patients may also be prescribed tablets to protect the stomach from side effects (eg. Ranitidine, Lansoprazole, Omeprazole or similar).

### **Azathioprine (Imuran)**

An important and very commonly used immunosuppressant. Major side effects are unusual but blood tests must be monitored frequently when it is started or the dose is increased because there is a risk of liver toxicity and bone marrow suppression (low white cell count and anaemia). Other side effects include fatigue, hair loss, diarrhoea, and increased risk of infection.

### **Methotrexate**

An important and very commonly used immunosuppressant. Major side effects are unusual but blood tests must be monitored frequently when it is started or the dose is increased because there is a risk of liver toxicity and bone marrow suppression (low white cell count and anaemia). Patients should also be aware that methotrexate can cause short and long term lung damage which should be monitored. Other side effects include: gastro intestinal (GI) upset, dyspepsia (indigestion), dizziness, fatigue, chills, headaches and mood changes.

### **Mycophenolate Mofetil (Cellcept) and Mycophenolic acid (Myfortic)**

An important and relatively commonly used immunosuppressant. Major side effects are unusual but blood tests must be monitored frequently when it is started or the dose is increased because there is a risk of bone marrow suppression (low white cell count and anaemia). Side effects include: diarrhoea, vomiting, dyspnoea (shortness of breath), insomnia, skin irritation and flu like symptoms. Stomach and bowel side effects are relatively common.

### **Ciclosporin**

An immunosuppressant which works by reducing the function of lymphocytes (a type of white cell). In the long term it can cause chronic damage to the kidneys and this should be carefully monitored. In high doses it can cause reversible problems to the kidney or liver function and cause paraesthesia (tingling in hands and feet), fatigue and headache.

**Tacrolimus** - Similar to Ciclosporin

### **Etanercept**

An artificial manufactured protein that can be injected to reduce the effect of tumour necrosis factor alpha (TNF). TNF is an important protein in the body that can cause inflammation and is important in fighting infection. Immediate side effects are unusual but can include: dyspnoea (shortness of breath), confusion, paraesthesia (tingling of the skin) and vertigo. A major side effect can be a reduced ability to fight some types of infection and possibly to control cancer cells in the body.

### **Immunoglobulin**

Normal human immunoglobulin is sometimes used to treat some forms of vasculitis, often when other treatments cannot be tolerated or have been ineffective. It is a particularly important treatment in Kawasaki's Disease. It is given as an intravenous infusion and repeated infusions may be necessary. Side effects are common during or

following each infusion including nausea, diarrhoea, chills, fever, headache, dizziness, joint, muscle and back pains. Rarely serious allergic reactions or kidney failure may occur.

### **Other commonly used drugs**

#### **Antihistamines**

Used to treat allergies and itching. Sedating antihistamines may initially cause sleepiness. Non-sedating antihistamines rarely cause drowsiness.

#### **Bisphosphonates (including Zoledronic acid, Risedronate and others)**

A major side effect of long term prednisolone use is osteoporosis. Bisphosphonates may be prescribed to strengthen bones and help prevent osteoporosis. Side effects from oral bisphosphonates are usually mild but include nausea, dyspepsia, diarrhoea or constipation and headaches. They cannot be used in patients with marked kidney damage.

#### **Colchicine**

Colchicine is an anti-inflammatory drug commonly used to treat attacks of gout. It is also sometimes used to treat some forms of vasculitis such as Behçet's disease and hypersensitivity vasculitis. It is usually given in a tablet form and often only for short periods. Common side effects are nausea, vomiting and abdominal pain. Patients with impaired kidney function may not be able to remove the drug properly from the body and short courses only should be prescribed. It rarely causes damage to kidneys, liver, nerves and muscles. Long term use should only be under expert supervision.

#### **Co-trimoxazole or Septrin (Bactrim)**

An antibiotic prescribed to some patients being treated with high doses of immunosuppression to prevent the lung infection pneumocystis carinii (also known as pneumocystis jirovecii) pneumonia. It is also sometimes used in patients with Wegener's granulomatosis (granulomatosis with polyangiitis) as it may reduce the risk of relapse in some patients. Side effects include: nausea, diarrhoea, headache, rash and rarely a low white cell count

#### **Dapsone**

An antibiotic which is sometimes used to control vasculitis limited to the skin (neutrophilic dermatoses). It can cause nausea, dizziness, headache and insomnia.

#### **Mesna**

Mesna is an "uroprotectant" used to prevent bladder damage during intermittent cyclophosphamide treatment. Side effects include: nausea, diarrhoea, headache, rash and dizziness.

#### **Non-steroidal anti-inflammatory drugs (NSAIDs) and Cox 2 inhibitors**

These groups of drugs includes ibuprofen, diclofenac (voltarol) indomethacin, celecoxib and etoricoxib. They are commonly prescribed for the control of pain, especially joint pain and can be very effective. Some of them are available without prescription. They should be used with caution as they commonly have side effects and can interfere with other prescribed medication. Even the drugs available without prescription can cause problems and patients should discuss with their medical team before taking them. Side effects are common with these drugs. Serious side effects include the risk of stomach ulcers and bleeding ulcers, an increase in the overall risk of bleeding and an increase in the risk of heart attacks and strokes. These drugs can also cause serious problems for patients with any kidney disease and they should not be taken by patients with kidney problems without careful discussion with the medical team. They should not be taken by patients with a history of stomach ulcers. They can also cause problems with fluid retention and high blood pressure.

## Glossary of some procedures undertaken in the treatment of vasculitis



**Note - Most of these procedures are undertaken generally in the diagnosis and treatment of illnesses and diseases, and are not specific to vasculitis. However, they may be used as part of diagnosis and treatment of vasculitis.**

### **Abdominal Ultrasound**

Uses sound waves to create a picture of the organs and structures in the abdomen. This test may show if abnormalities are present in the abdomen.

### **Angiography**

Angiography is a way of looking at the blood vessels either to identify blockages, inflammation or other abnormalities. There are many ways of performing angiography. Ultrasound can be used to look at large vessels without needing to use x-rays or dyes. Magnetic resonance imaging (MRI) uses magnets to create detailed images of some blood vessels and may or may not need special dyes (contrast agents) to be injected into the patient. Computed tomography (CT) scanning uses x-rays to look at the blood vessels and usually needs contrast dyes to be injected. Positron emission tomography (PET) scanning requires the injection of radioactive drugs into the patient and may be helpful in locating areas of blood vessel inflammation. The best test to use depends on the clinical question that the medical team need answering.

### **Angioplasty**

A treatment for narrowed, scarred or "furred up" blood vessels. It involves widening one or more of the narrowed arteries to allow blood to flow more easily through to the organ or limb it supplies. Stenting may also be performed as part of angioplasty. This is where a narrow metal tube is inserted into the blood vessel at the site of the narrowing to help keep the blood vessel open and prevent the narrowing recurring.

### **Anticoagulants (INR test)**

For patients taking anticoagulant (blood thinners) eg Warfarin. (INR = International normalised ratio). A system for reporting the results of blood coagulation (clotting) tests.

### **Biopsy**

A procedure whereby a small piece of tissue is taken from the affected area or organ, eg the skin or the kidney. The tissue is then examined by a pathologist and special tests can be done to identify the disease affecting the organ.

### **Blood pressure monitoring**

Should be monitored in all cases of vasculitis. Elevated blood pressure in vasculitis can lead to kidney damage.

### **Bronchoscopy**

A procedure where the physician looks into the large airways (trachea and bronchi) and identify areas of damage or collect samples (biopsy or washings) to identify the disease or infection. These are the main tubes that carry air into the lungs. A fibre-optic bronchoscope is usually used (a thin, flexible, telescope).

### **Computerised tomography (CT scan)**

Gives greater detail of the internal organs than a normal radiograph (x-ray).

### **DEXA scan (Dual energy x-ray absorptiometry)**

A test that measures the density of bones. This can be used to identify or monitor the strength of bones particularly for patients on long term steroid medication.

### **Dialysis**

When the kidneys do not work adequately toxins, acid and fluid accumulate in the body. The process of dialysis helps removes this from the body to keep patients with severe kidney failure alive. Sometimes this treatment is only needed temporarily (days, weeks or months) to give time for the kidneys to recover where the cause of kidney damage is treatable. Where the kidney damage is not reversible dialysis treatment is permanent. For patients needing long term dialysis this can be done at home or as an outpatient.



### **Echocardiography**

An ultrasound test using sound waves to create pictures of the heart. The test will indicate the size and shape of the heart and how the chambers and valves are working.

### **Electrocardiogram (EKG) (ECG)**

A simple and painless test to record the heart's electrical activity. This can provide useful information about the state of the heart and can give indications if the heart is affected by vasculitis.

### **Electromyography (EMG)**

This is an electrical stimulation test of the muscles (usually in the arms or legs) to look for evidence of abnormal muscle function such as can be seen in vasculitis. Often it is performed in conjunction with a nerve conduction test.

**Lung function tests** – see Spirometry



### **Magnetic Resonance Imaging (MRI)**

Uses magnets and a computer to create detailed pictures of the internal organs.

### **Plasma exchange or plasmapheresis**

This treatment is sometimes used in patients with severe vasculitis where antibodies in the blood are thought to be important in causing the disease. The treatment involves removing antibodies from the blood using a machine and returning the "cleaned" blood back to the patient. The treatment may necessitate giving blood products to the patient including plasma, albumin or immunoglobulin. It may also involve giving drugs to thin the blood and prevent it clotting in the machine.

### **Positive emission tomography (PET scans)**

Produce 3-dimensional pictures which show the level of metabolic activity - and indicator of inflammation.

### **Spirometry (lung function test)**

Measures the size of the lungs, the amount of air inspired and expired and how well the lungs deliver oxygen to the blood. This can be useful for monitoring patients whose lungs or airways have been affected by vasculitis.

### **Tracheostomy**

Tracheostomy is sometimes necessary for patients whose main airway has become blocked or narrowed as a result of damage caused by vasculitis. It provides direct access to the trachea, bypassing the blockage, by surgically making an opening in the neck. This allows air to enter the lungs and permits patients to breathe properly. Tracheostomy can be either a temporary measure, particularly for patients who are severely ill on a ventilator, or a permanent measure where irreversible damage has occurred. Patients with permanent tracheostomies have a tube to keep the opening open and allow normal speech and otherwise live a normal life.

**Ultrasound** (see Abdominal Ultrasound)

### **Urinalysis**

A simple but very important test of the urine, commonly undertaken using "dipsticks", which identifies abnormal levels of protein or blood cells in the urine. Abnormal levels of protein or blood can be a sign of kidney involvement.

### **Vaccines**

Vasculitis patients who are immuno-compromised should **not** receive live vaccines. Influenza and Pneumonia vaccines are not live vaccines and **are** recommended.

## Glossary of Blood Test Monitoring

**Interpretation of blood test results** Normal ranges differ between different hospital laboratories and may not be comparable. Therefore, blood results should be discussed with the medical team.



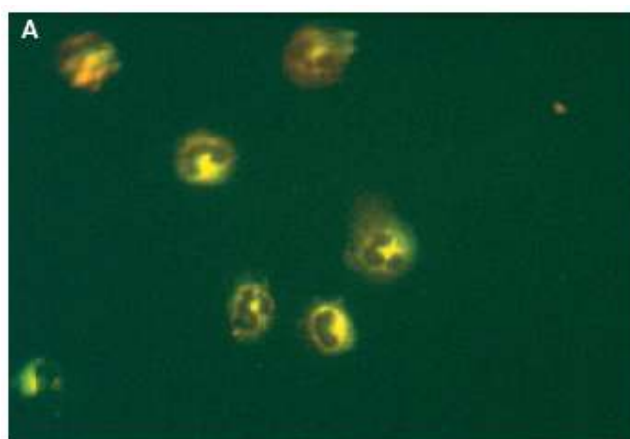
### **Alkaline phosphatase (Alk. Phos):**

Alkaline phosphatase is an enzyme made in the liver, bone, and the placenta and normally present in high concentrations when during growth. Alkaline phosphatase is released into the blood during injury and during bone growth and pregnancy. Abnormally high blood levels may indicate disease, eg in the liver or bones.

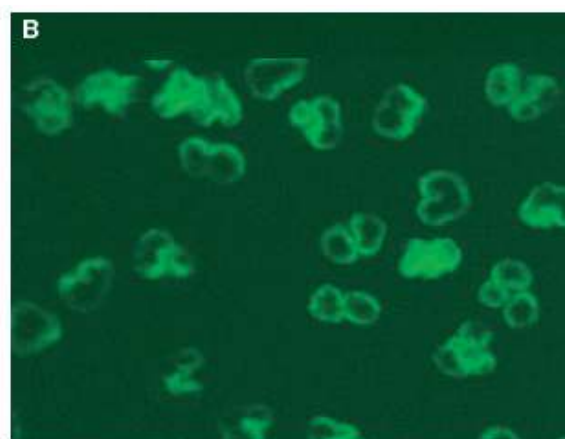
### **Antineutrophil Cytoplasmic Antibodies (ANCA):**

Autoantibodies which are usually found in patients with the ANCA associated vasculitides but can also be found in other inflammatory diseases (particularly inflammatory bowel disease), some infections and occasionally in entirely healthy people.

There are two main types of ANCA found in people with vasculitis called c-ANCA (which is usually the same as anti-proteinase 3 or anti-PR3 ANCA) and p-ANCA (which is usually the same as anti-myeloperoxidase or anti-MPO ANCA). One of the main tests for ANCA involves looking down a microscope at neutrophils to see where the ANCA is sticking to the neutrophil. Using this test the c-ANCA and p-ANCA look different as in the pictures below.



c ANCA



p ANCA

Images courtesy of Dr Lisa Willcocks, Addenbrooke's Hospital, Cambridge

c-ANCA is more commonly found in patients with Wegener's granulomatosis (also called granulomatosis with polyangiitis) and p-ANCA is more common in microscopic polyangiitis although either can be found in both diseases. People with Churg-Strauss Syndrome may have p-ANCA or c-ANCA or no ANCA at all.

### **Aspartate Aminotransferase (AST):**

AST is an enzyme found in heart muscle, liver and skeletal muscle cells. To a lesser extent it is found in other tissues. Raised levels can be found following damage to the heart, liver or muscles.

### **Creatinine:**

Creatinine is measured in the blood as a measure of kidney function. Creatinine is released into the blood mainly from muscle cells and is removed by the kidneys. In general the higher the creatinine level in the blood the worse the kidney function is. Creatinine has several serious limitations for measuring kidney function. It varies hugely from patient to patient even when the kidney function is the same. The level in the blood is also dependent on the age, gender, body size, muscle mass and diet of the patient. It is also possible to have normal creatinine levels in the blood with very abnormal kidney function. Sometimes it is also possible to have abnormal creatinine levels with normal kidney function.

Because of these limitations most doctors now measure kidney function as the estimated glomerular filtration rate (eGFR) which can be more reliable.

**C-Reactive protein (CRP):**

C-reactive protein is produced by the liver. The level of CRP usually rises when there is inflammation present although this is not specific for vasculitis and can also indicate infection or other problems

**Erythrocyte sedimentation test (ESR):**

ESR is a test that indirectly measures how much inflammation is in the body.

**Haemoglobin (Hb):**

Haemoglobin is a protein in red blood cells that carries oxygen. Measuring the Hb level is useful to detect a low red blood cell count (anaemia).

**Platelets:**

Platelets assist in blood clotting. During normal blood clotting, the platelets clump together. Although platelets are often classed as blood cells they are more accurately fragments of large bone marrow cells called megakaryocytes. The platelets are usually measured as one of the measures of bone marrow function. Very low levels can increase the risk of bleeding. High levels can occur if inflammation is present.

**White blood count (WBC):**

White blood cells (also called leukocytes) are an important part of the immune system. The cells help fight infections and also monitor the body for the development of early cancer cells which they destroy. They also have an important role in the inflammation which occurs in vasculitis which leads to organ damage. There are five major types of white blood cells: Basophils, Eosinophils, Lymphocytes (T and B cells), Monocytes, and Neutrophils which all have different roles in the body. Monitoring the WBC is important for patients on immunosuppressant drugs as a low WBC can be caused by many of these drugs and can lead to infection if the immune system is over suppressed.

## Glossary of genetic terms

A glossary of genetic terms (not necessarily related to vasculitis) can be found at the Genetic Alliance UK website: [www.geneticalliance.org.uk/glossary.htm](http://www.geneticalliance.org.uk/glossary.htm)

## Living with Vasculitis

### General Guidance

The nature of vasculitis together with the immunosuppressive treatments often make it difficult for a newly diagnosed patient to know whether a particular medical problem or side effect is due to the disease, the drugs used, or another totally separate condition. Patients should, therefore, educate themselves about their particular vasculitic disease, including medication, side effects and potential problems.

Eventually you will become aware of what is and is not normal in your individual case.

Managing vasculitis is very much a team effort between you and your physician. The patient who is informed and educated about their condition can sometimes help the physician by asking the appropriate questions about their diagnosis and ongoing treatment.

Over time you will become more aware of what is and what is not normal in your own case. When a patient is unsure about any aspect of their condition or treatment, particularly whether a problem might be due to the medication or even to a possible relapse, it is important to contact the physician and not to wait until the next hospital appointment.

Vasculitis treatment invariably is undertaken by a hospital physician and not by the GP as the latter do not have expertise in vasculitis treatment. The hospital physicians who deal with vasculitis include: Rheumatology, Nephrology (kidney), and Ear Nose and Throat) although others may also be involved. Knowledge and expertise varies widely throughout the UK. Should you be unhappy with any aspect of your treatment you should not be afraid to seek a second opinion.

### Understanding feelings and reactions to illness

There are many feelings associated with illness, some may not have been experienced before or may be more intense than previously encountered. These may include frustration, anger, anxiety, fear, vulnerability, and even being ashamed of being ill. Some patients feel a sense of loss and grief and may even deny the fact that they are ill. Feelings of uncertainty also occur quite frequently during a time when so much change is being experienced. Tearfulness at times, depending on personality, is common.

All these feelings are to be expected and are part of the normal process of dealing with and adapting to a major life changing episode. It often takes time to come to terms with change. This may be weeks, months or even years, depending on how many and what types of changes have taken place. Just as it takes time to get over the loss of a loved one, so it takes time to get over the loss of a way of life or good health. The ongoing support and understanding of family and friends can be of great help during such distressing times. It is often helpful to accept this support, even when this may be alien to the patient's nature.

For vasculitis patients suffering from low mood or depression Manchester NHS has produced a booklet "**Improve your mood: A guide to managing low mood and depression**" - see "Low Mood and Depression " [www.vasculitis.org.uk/living-with-vasculitis/depression](http://www.vasculitis.org.uk/living-with-vasculitis/depression)

It is also important to remember that not only you, the patient, feels the loss. Family and friends are also affected. Talking about feelings and difficulties can be helpful and may lead to coping better with the difficulties and prevent misunderstanding.

One of the problems of having a rare disease is that family, friends and work colleagues often find difficulty understanding what is happening - particularly if symptoms are hidden and the patient 'looks well'.



As a patient with a rare disease you may find that the concept of your problem is not easy to grasp by other people. Perseverance, whilst frustrating, is helpful in raising awareness.

Where the vasculitis has resulted in limitations of any kind, you may experience negative attitudes in others. Patience and dialogue may be required.

Family and friends may find the "Caring for a family member with vasculitis" section helpful reading.

Many vasculitis patients comment that they have lost their "old life". They may no longer be able to participate in their favourite activities or interests. The "new life" might not be what you would have envisaged but it is necessary to change and adapt. Taking up a different hobby or using skills previously acquired in a different endeavour can be helpful.

## **Pain Management**

### **General Information**

Discussion with the GP or consultant is important when experiencing pain. Where the drug regime prescribed does not have beneficial results this should be reported. Often the drug dosage may simply require alteration.

When one drug does not work the physician may prescribe something different.

Over-the-counter drugs should be avoided when taking prescribed medicines as used in the treatment of vasculitis. Advice is available from the pharmacist.

For mild to moderate pain paracetamol and rest, at the onset of the pain, are recommended. Fear, anxiety and stress can exacerbate pain.

Simple relaxing techniques can be beneficial. For severe pain the GP or consultant may recommend referral to a Pain Management Centre.

### **Managing Chronic Pain**



The Scottish Intercollegiate Guidelines Network (SIGN) writes guidelines which give advice for healthcare professionals, patients and carers about the best treatments that are available. These guidelines are written by working with healthcare professionals, other NHS staff, patients, carers and members of the public. The guidelines are based on the most up-to-date scientific evidence.

SIGN produced a booklet for patients and carers "Managing Chronic Pain" in December 2013. They have very kindly given Vasculitis UK permission to reproduce this excellent booklet. [www.vasculitis.org.uk/living-with-vasculitis/dealing-with-pain](http://www.vasculitis.org.uk/living-with-vasculitis/dealing-with-pain)

The booklet contains information on: What is chronic pain? Those involved in patient care. How pain is assessed. What to do to help yourself. Medication available to manage pain. Physical therapies and other methods to help manage pain. The roles of complementary medicines and diet.

### **Low Mood and Depression**

Many patients with long term illness suffer from some degree of low mood or depression. This is a normal phenomenon and is often transient.

When a problem persists this should be discussed with your GP or consultant. A short course of anti-depressants may be prescribed. Occasionally counselling may be indicated.

### **Improve your mood: A guide to managing low mood and depression**

Manchester Mental Health and Social Care Trust, has produced a guide [Improve your mood](http://www.vasculitis.org.uk/living-with-vasculitis/depression) which covers a number of different techniques to help you manage low mood and depression. The guide can be downloaded at: [www.vasculitis.org.uk/living-with-vasculitis/depression](http://www.vasculitis.org.uk/living-with-vasculitis/depression)

The techniques are provided under the headings of "See it", "Treat it" and "Beat it". It is recommended that you try one of these ideas as a way to start. You can then move on to try some of the others at your own pace.

## **General Guidelines**



You may be the only vasculitis patient on your GP list. Where possible you should arrange to see the same doctor at each visit to ensure continuity of treatment and advice given.

Many vasculitis consultants advise patients to contact them (via their specialist nurse, secretary or clinic) regarding new symptoms and problems encountered as a result of changes in medication.

As you become more educated in the disease process you will understand what is normal in your particular case. However, it is often difficult to detect whether any change in condition is due to the onset of a flare or relapse, or

whether it may be attributable to a simple infection. Consultation is advisable under these circumstances

**Hospital Consultations:** As part of the consultation process, the physician will discuss the findings and ask general questions. It is important to take a written note, as an aide memoire, of matters which need to be brought to the attention of the physician. Where you do not understand any aspect of what the doctor has said then clarification should be sought. Taking a family member or friend (an advocate) into the consultation can be very helpful in enabling you to recall any important instructions given. Many patients find it useful to keep a diary of main events, symptoms and other key aspects of their vasculitis - referring to past entries can help prepare a written note for the consultation. It also serves to show that progress is being made.

Patients may not wish to mention a particular problem they may be encountering mistakenly thinking it is of no importance or they fear that the medication or treatment regime may be increased or changed. The physician requires the full facts in order to provide appropriate care.

Changes to treatment regimes are not made solely on test results. The physician will consider, amongst other things, recent blood, urine and other laboratory tests, what they find examining the patient and the history given. Blood tests are only a tool and a guide for the physician. For example, a negative ANCA test does not automatically mean the disease is quiet, whereas a positive ANCA does not automatically mean the patient is not responding to treatment.

When seeing the doctor you should never be afraid to ask any questions regarding your disease, possible symptoms or treatment. The doctor should be happy to discuss any changes to treatment to enable you to make an informed decision and to reach agreement with you about what is appropriate in the individual case. Where you are not happy with the explanations given, suggested treatment or changes in treatment you should not be afraid to say so and ask for clarification.

If you are seriously dissatisfied with the treatment received you should discuss this with the consultant. If you are still dissatisfied you should ask your GP to refer you to another consultant/hospital. Under current NHS regulations regarding "Patient Choice" patients are entitled to be referred for a second opinion to a consultant or hospital of their choice. The request to be seen by another consultant cannot be over-ruled by the consultant concerned. If the GP does not co-operate the patient should ask for a written letter explaining why the request has been declined.

## Regular Blood Monitoring

Even when the disease is in remission (quiet) it is necessary to have blood tests undertaken at regular intervals. Initially blood monitoring may be undertaken weekly or fortnightly. Depending on the type of drugs prescribed the periodicity of blood monitoring will vary. It will normally be every month or every three months.

## Helpful Hints

**Balanced lifestyle** - You should adopt a balanced lifestyle remaining aware of your individual body's ability. It is important to rest and not become exhausted. Resting before becoming over-tired will help maintain a steady obtainable pace and reduce the changes of requiring an enforced rest.

**Rest and steady exercise** - Rest and steady exercise are important components in the recovery process. These will ensure muscles remain strong and flexible.

**Diet** - Healthy well balanced food intake is important since too much will cause weight gain in addition to that experienced by the use of steroids. (See separate section on Diet below).

**Diabetes** - Some patients may also become diabetic. This is controllable with appropriate drugs assisted by a healthy diet.

When a diabetic patient is diagnosed with a disease requiring steroids, short-term insulin might be required to control the blood sugars.

Basic information on diet can be found in this publication. Information is available in most G.P's surgeries and also in food stores. Some forms of vasculitis or treatment require special diets and these should be discussed with a dietician. You should speak to your GP for referral to a community dietician.

**A Positive attitude** - Having a positive attitude always helps with vasculitis as does trying to maintain a cheerful disposition. Planning to achieve something each day and noting the level of activity in a diary can indicate how much improvement is being achieved on a day-to-day basis.

### Useful hints for:

Congested Nose/Sinuses - Steam inhalation and nasal doused with Betnessol N nasal drops then Glycerin and Dextrose Nasal drops

Dry, flaky skin - Lotion or Cream, e.g. E45

Mouth Ulcers - Bonjela

Stiffness, aches and pains - warm baths with Radox or Radium B

Deafness - Ears can sometime be "popped" by chewing gum.

## Diet

A healthy diet is important for the vasculitis patient, especially for those who are struggling with food intake. Healthy eating will, by definition, help anyone who has diabetes or who have gained weight and wants to do all they can nutritionally to stave off infection and disease long term.

Some vasculitis patients will require a special diet and this will be arranged via the hospital dietician. It is important to adhere to this regime.

For those with kidney involvement you may find the kidney care cookbook **Rediscovering Food and Flavours** from Kidney Research UK helpful. This cookbook was created by TV Chef Lawrence Keogh, Head Chef at Roast and BBC's Saturday Kitchen along with Renal Dietician, Diane Green. The book contains 16 recipes. It is now available free of charge from local Dieticians for patients who need to control their diet due to chronic kidney disease. It can be downloaded at:

[www.kidneyresearchuk.org/health-information/resources/free-recipe-book](http://www.kidneyresearchuk.org/health-information/resources/free-recipe-book)

If you do not need a special diet, you should aim to cut down on starchy foods - bread, potatoes, rice and pasta, replacing these with fresh fruit and vegetables. You should also avoid processed food and grain fed meat. The



omega 3 fats in oily fish such as salmon, mackerel, trout and sardines are beneficial in autoimmune disease. Also omega 3 can be found in flaxseed, walnuts and green leafy vegetables.

Omega 3 fish oil supplements containing EPA and DHA can be helpful. However, it should be noted that these do react with some medication. These, and other supplements, should not be taken without discussion with your doctor or nutritionist.

A sensible eating regime should be adopted especially for those patients taking steroids. This will help control weight gain. Excessive dieting is not recommended for the vasculitis patient. Also, when taking steroids there is an increased risk of developing osteoporosis. Increased calcium in the diet can help prevent osteoporosis developing. Eating broccoli, yogurt, skimmed milk and tinned sardines are recommended.

Where the drug regime allows the drinking of alcohol this should only be in moderation. There are some immunosuppressant drugs where drinking alcohol is contra-indicated. Your doctor will discuss this with you if it is relevant in your case.

## Vasculitis and Oral Health

Vasculitis can affect all parts of the body and the mouth is no exception. Large, persistent and excruciatingly painful mouth ulcers are sometimes a characteristic of active Wegener's Granulomatosis as is severe toothache that moves around the mouth, especially in the upper jaw.



When patients are severely ill with vasculitis (or any other disease) it is essential that their dental care is not neglected, as a period of neglect can cause permanent damage to teeth and gums. This can also apply where patients are suffering from restricted use of their hands or upper limbs.

Obviously, those taking immuno-suppressing drugs must take care as the mouth is one of the places where invading bacteria can most easily gain the upper hand.

Some medication, especially some types used to control high blood pressure, can have the side-effect of making the gum tissues swollen so that they bleed and are more prone to infection.

Pre-existing dental disease, in the teeth or gums, may get worse due to steroids or immuno-suppression, so these need immediate attention by the dentist.

The vasculitis patient should tell their dentist that they have vasculitis. Also if the patient is taking steroids, immunosuppressing drugs or drugs to prevent osteoporosis, they **must** tell their dentist. The drugs used to counteract osteoporosis, such as the bisphosphonates or alendronates, can cause serious problems if a tooth has to be extracted.

Like in vasculitis generally, damage once done, cannot always be reversed. The best remedy, as usual, is prevention.

- 1) The **proven** most effective tooth brushing regime is to use an electric toothbrush with a rotary oscillating head, such as the Braun Oral B
- 2) Couple this with a toothpaste containing both fluoride and triclosan, such as Colgate Totalcare
- 3) If possible use dental floss or dental tape regularly or interdental brushes
- 4) For gum problems, use an antiseptic mouthwash containing a low dose of chlorhexidine, such as "Corsodyl". This may be used daily for a short term, but in the long term, only as a weekly mouthwash as it can cause staining of the teeth, although this is easily removed.



## Vaccinations

Patients taking immune-suppressants should not receive live vaccines such as yellow fever, typhoid and measles. Live vaccine contains a small amount of the infection itself. Under normal circumstances the patient can fight off this small amount of infection but this is not possible for the vasculitis patient. This will have implications for holidays in destinations and where live vaccines are recommended, eg Africa, Asia, and Latin America.

Flu and Pneumonia vaccines **are recommended** for vasculitis patients. These are **not** live vaccines. Where the patient is unsure about a particular vaccine it is important to speak to the doctor about this issue.

For any other types of vaccination it is essential to question the need or reason for these. You should ascertain what they contain and discuss any concerns with the GP.



## Fertility and Vasculitis



The Trust is indebted to Dr David Jayne, Vasculitis and Lupus Clinic, Addenbrooke's Hospital, Cambridge, UK, for providing the following information on fertility and vasculitis:

### **Introduction:**

Vasculitis activity and its therapy are potential threats to the fertility of patients with vasculitis. Loss of fertility is an important consequence of the disease, but the risks of this occurring can be considerably reduced with newer forms of treatment. As a chronic disease, vasculitis also causes psychosexual and relationship problems due to effects on self-esteem and mental well-being. Chronic kidney disease is a common consequence of renal vasculitis

and the depressed kidney function itself affects fertility in both women and men.

### **Fertility in women:**

The major threat to women is cyclophosphamide exposure. This drug is used to control vasculitis activity and is directly toxic to the ovaries. This can result in permanent infertility, also known as primary ovarian failure. In addition to loss of menstrual periods, amenorrhea, blood levels of the hormones FSH and LH are elevated. Primary ovarian failure is related to the total amount of cyclophosphamide administered and to the age of the patient. Data from lupus nephritis suggests that a total cyclophosphamide exposure of 14-20g results in infertility in over 50% of women aged over 32 years. The risk of infertility in those under 32 years is lower, around 10% in one series. These risks can also be reduced by using short-term regimens or by intravenous pulse as opposed to daily oral administration. Three months of oral cyclophosphamide leads to an exposure of 9-14g, and an equivalent six dose course of intravenous pulse cyclophosphamide, 5-7g. Even if infertility is not induced, less severe ovarian damage leads to earlier menopause. Drugs that temporarily suppress ovarian function, such as zolodex, are used to reduce the risk of cyclophosphamide toxicity. Rituximab has been shown to be as effective as cyclophosphamide and can be used when cyclophosphamide avoidance is desirable.

Following cyclophosphamide withdrawal and return of a normal menstrual cycle, women can conceive and have children. There have been concerns that cyclophosphamide, through damage to DNA in the unfertilised egg, results in an increase in birth defects but this has not proved to be the case. However it is advisable to wait at least six months between stopping cyclophosphamide and attempting to conceive.

Women have a finite number of eggs and once lost, they cannot be replaced. For this reason, in cancer therapy, egg harvesting and preservation or even, preservation of ovarian tissue for subsequent re-implantation, can be considered when drugs toxic to the ovaries are used. This is rarely feasible in vasculitis due to the tempo of the disease, the need for rapid institution of therapy and the potential complications of the procedures.

Being unwell with vasculitis interferes in a non-specific manner with menstrual cycles and can cause periods to stop, amenorrhea. This can be differentiated from ovarian damage by an ultrasound scan of the ovaries which demonstrates the presence of healthy oocytes (egg follicles) and by measuring hormone levels in the blood. Periods will usually re-start spontaneously as the patient recovers. Direct damage to the ovaries or female reproductive tract by vasculitis is rare but has occurred.

The influence of other drugs should also be considered. Some immune suppressives, such as, methotrexate and mycophenolate mofetil, and thalidomide, damage the foetus and must not be used in pregnant women or those attempting to conceive. Certain antibiotics, such as rifampicin, interfere with the contraceptive pill. Anti-inflammatory drugs and high dose steroids also reduce fertility. The infective risks of the coil are increased in those receiving immune suppression. Sexually transmitted diseases can be more problematic in immune suppressed patients and *Chlamydia Trachomatis* can result in infertility in women.

### **Fertility in men:**

Cyclophosphamide directly affects sperm production in men but there is more potential for recovery by the generation of new sperm forming cells when cyclophosphamide is withdrawn. However sperm production does not usually recover to pre-treatment levels and healthy sperm counts can remain depressed. It is likely that, in

combination with non-specific effects of chronic illness, cyclophosphamide reduces male fertility. An alternative immune suppressive used in vasculitis, methotrexate, also reduces sperm formation but has a lower risk of sustained effects after drug withdrawal.

In contrast to the difficulties of egg preservation in women, semen preservation is quite feasible in men and can be considered before cyclophosphamide is commenced. It can be hard to advise on this, especially if there is a cost to the patient involved, because the risks of infertility are probably quite low with current cyclophosphamide regimens and if infertility occurs it is more likely to be partial rather than complete. Testosterone therapy has also been used to protect the testes from cyclophosphamide toxicity.

The testicles can be directly attacked by vasculitis in polyarteritis nodosa causing pain and swelling and subsequent loss of function. GPA (Wegener's) can affect the prostate gland but the effects on fertility are not well understood.

Drug effects, especially high dose steroids, vasculitic activity and chronic illness reduce testosterone levels that can lead to reduced libido and erectile failure. Testosterone levels in the blood are readily measured and testosterone supplementation can correct the problem.

#### **Conclusions:**

The protection and preservation of fertility are important issues in the management of vasculitis. Much is known about the effects of cyclophosphamide in women although there is a shortage of information about the longer-term effects in men and in children. The potential dangers of this and other drugs should be discussed in detail before their use, but it should be remembered that cyclophosphamide has been a truly life saving drug in vasculitis and a balanced approach to assessing both risks and benefits is needed.

Many other factors will influence the fertility of vasculitis patients that include the consequences of vasculitic activity, ongoing medications and the patient's general state of health. Pre-conception counselling should be sort by women with vasculitis wanting children so that any risks to the mother, pregnancy or future baby can be discussed and appropriate plans made.

***Information of fertility and contraception will be available from your GP or Practice Nurse. There are a number of assisted conception units throughout the country. It is possible to do an internet search for these under "assisted conception" and add the name of the city. However, further information in the form of a booklet "Birmingham Women's Fertility Centre - A Patient Guide" is available from the Birmingham Women's Fertility Centre. Contact is by completing the web form at: [www.bhamivf.com](http://www.bhamivf.com)***

## Vasculitis and the Eye

The Trust is indebted to Dr Catherine Guly, Consultant Ophthalmic Physician, Bristol Eye Hospital for providing the following information on vasculitis and the eye

Vasculitis can affect different parts of the eye. In some patients there is only mild inflammation which does not affect the vision and in others symptoms are more severe and the vision may become affected. With advances in the understanding of ocular inflammation it has become clear that the treatments for vasculitis elsewhere in the body are also useful for treating vasculitis affecting the eye. This article discusses the different forms of vasculitis and explains how they can affect the eye.

### How is vasculitis of the eye diagnosed?

Vasculitis affecting the eye is usually diagnosed by an ophthalmologist (eye doctor). Ophthalmologists use a slit lamp which has a microscope that gives a magnified view of the eye. It is possible to look into the back of the eye by enlarging the pupil with dilating eye drops.

The eye examination can tell which part of the eye is inflamed but does not show what has caused the inflammation and so other investigations, such as blood tests, can be helpful in making a diagnosis. Blood markers of inflammation (including the CRP, ESR and plasma viscosity) are useful in diagnosing and monitoring giant cell arteritis. If there is inflammation behind the eye a CT or MRI scan can be useful. Rarely, a small sample of tissue is taken from the eye or around the eye to send for analysis in the laboratory to look for signs of vasculitis. In Giant cell arteritis a sample of artery from the temple (the temporal artery) is used to help with the diagnosis.

### How do different forms of vasculitis affect the eye?

**Giant cell arteritis (temporal arteritis)** can result in an optic neuropathy in one or both eyes. An optic neuropathy is a disruption of the function of the optic nerve, in this case due to inflammation of the arteries that supply blood to the optic nerve. The optic nerve joins the eye to the brain.

Patients with an optic neuropathy due to giant cell arteritis may notice loss of vision or transient loss of vision in one or both eyes. This is often associated with a headache. There may be pain on eating if the blood supply to the jaw is also affected. Early treatment with steroids usually stabilises the vision but if there is damage to the optic nerve it does not always recover. Occasionally, giant cell arteritis disrupts the blood supply to the retina (retinal artery occlusion) or results in double vision (cranial nerve palsy).

**ANCA associated vasculitis (Wegener's granulomatosis and Microscopic polyangiitis) and Polyarteritis nodosa** can affect all the different parts of the eye. The most common symptoms of vasculitis are redness and eye pain. There may be increased sensitivity to light.



Some forms of inflammation are mild and do not affect the vision. For example, episcleritis is the inflammation of the outside coat of the eye, the episclera, and may result in a red, irritated eye. Other forms of inflammation are more serious and may affect the vision if untreated. These include inflammation of the cornea which is the clear window at the front of the eye (keratitis), inflammation inside of the eye (uveitis) or inflammation of the sclera which is the white outer coat of the eye (scleritis). Scleritis in particular may cause severe pain around the eye, although this usually settles with treatment.

Orbital inflammation is inflammation in the eye socket. With orbital inflammation there may be double vision, the eyelid may be higher or lower than normal and the eye may protrude more than normal. The vision can become

blurred if the optic nerve is affected. Orbital inflammation is most commonly associated with Wegener's granulomatosis.

**Churg Strauss syndrome** rarely causes inflammation in the eyes.

### **How is vasculitis of the eye treated?**

The treatment will depend on the type of vasculitis and which part of the eye is inflamed. Eye drops can be used to treat inflammation at the front of the eye such as episcleritis and some forms of uveitis. Other types of inflammation usually require high dose steroid treatment with or without other immunosuppressive medications (such as mycophenolate mofetil, cyclophosphamide or methotrexate) or biological medications (such as adalimumab, infliximab and rituximab). Giant cell arteritis can usually be treated with steroids alone but sometimes other immunosuppressive medications are added. Patients usually require high doses of treatment initially to gain control of the inflammation and then the treatment is tapered to the lowest dose that will maintain control of the inflammation.

If the patient has inflammation elsewhere in the body the ophthalmologist will aim to work closely with the other physicians involved so that as far as possible the medicines chosen will treat the inflammation in the eye as well as any inflammation elsewhere in the body. The treatment will normally be dictated by the organ which is most at threat so if the eyes are inflamed and the sight is at threat then the ophthalmologist will need to guide the type of treatment required and how quickly the treatment can be reduced.

### **Is there anything I can do to protect my vision?**

If you have vasculitis you should report any new visual or eye symptoms to your doctor or your optometrist (optician), particularly if you notice any change in your vision. It is also a good idea to see an optometrist (optician) on a yearly basis as they will perform a thorough eye examination during your eye check. An optometrist can also detect any side effects from long term use of prednisolone (steroids), such as cataracts and more rarely, glaucoma.

### **More detailed medical information about how vasculitis affects different parts of the eye**

All the conditions detailed here may be associated with vasculitis but are also found with other medical conditions.

**Conjunctivitis** is inflammation of the conjunctiva. When the conjunctiva is inflamed the eye and inside the eyelids become red and the eye may feel gritty. The vision is usually unaffected. Conjunctivitis is also very common in the general population and is usually caused by infection or allergy, but may also be caused by vasculitis.

**Episcleritis** is inflammation of the episclera, which is the thin covering over the sclera. With episcleritis the eye looks red, and this may affect just a patch of the episclera so that just part of the eye is red, or it may affect the whole episclera. The eye may feel irritated and uncomfortable but the vision is unaffected. Episcleritis is diagnosed with a slit lamp examination. Phenylephrine eye drops blanch the affected area (unlike scleritis where the inflammation is deeper) and this can be helpful in making a diagnosis.

**Peripheral ulcerative keratitis** is inflammation and ulceration of the cornea. The eye is usually red and painful and the vision may be blurred. The inflammation and ulceration start at the edge of the cornea and in severe cases the cornea may perforate. Keratitis is diagnosed with a slit lamp examination. Corneal ulcers are detected using fluorescein eye drops; the epithelial defect takes up the stain and the ulcer glows yellow under a blue light.

Scleritis is inflammation of the sclera. When the sclera becomes inflamed the eye usually becomes red and painful. The pain often disturbs sleep and sometimes the vision becomes blurred. Occasionally, only the back part of the sclera becomes inflamed and this is called posterior scleritis. The posterior sclera is not visible from the outside of the eye and in patients with posterior scleritis the eye may be painful but does not look red. Scleritis is diagnosed using a slit lamp examination. An ultrasound scan of the eye is useful to detect posterior scleritis.

**Uveitis** is inflammation inside the eye. The 'uvea' is the pigmented layer inside the eye made of the choroid, ciliary body and iris, but uveitis also affects other parts of the eye like the retina and so it is easier to think of it as 'intraocular inflammation'. Uveitis can be divided up into anterior uveitis which affects the front chamber of the eye, intermediate uveitis which affects the middle part of the eye (the vitreous) and posterior uveitis where the back of the eye (retina and/ or choroid) is inflamed. In panuveitis the inflammation affects the front, the middle and the back of the eye.

Uveitis usually causes a red painful eye which is worse in bright light but in some forms of uveitis the eyes are white and there is no pain. If there is inflammation in the vitreous there may be multiple floaters that obscure the vision and if there is inflammation in the retina or choroid the vision may be blurred or there may be loss of vision.

The diagnosis is made using a slit lamp examination, and additional imaging of the retina and choroid (using fluorescein angiography, indocyanine green angiography and optical coherence tomography imaging) is helpful in some cases.

**Retinal artery occlusion** is where there is a disruption of the blood supply to the eye. This is rare in vasculitis but it results in sudden loss of vision. Further investigations are usually required to determine if the occlusion is primarily due to vasculitis or if there may have been an embolus (clot travelling to the artery from elsewhere in the body) resulting in the occlusion.

Optic neuropathy is an abnormality of the function of the optic nerve. Patients with optic neuropathy usually notice loss of vision, which can be sudden. Sometimes patients have transient vision loss. Patients may be aware of blurred or patchy vision and may notice that colours seem washed out. An optic neuropathy is diagnosed through tests of optic nerve function, including visual acuity, colour vision, visual fields and pupil examination. The optic nerve may appear swollen or pale. Other investigations which may be helpful include electrophysiology (to measure optic nerve function), MRI/ CT scan of the head and orbits and a lumbar puncture.

**Orbital inflammation** is where there is inflammation in the eye socket, which includes the muscles around the eye. With orbital inflammation there may be double vision, the eyelid may be higher or lower than normal and the eye may protrude more than normal. There may be pain or discomfort around the eye and if there is any pressure on the optic nerve there may be an associated optic neuropathy. The eyelids and conjunctiva may also be inflamed.

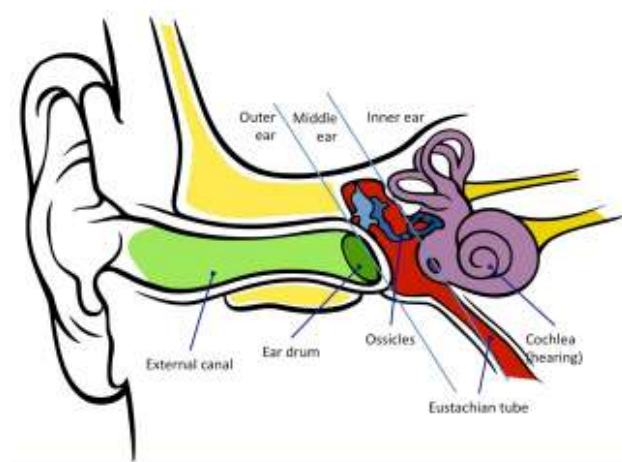
**Cranial nerve palsies.** Vasculitis can disrupt the blood supply to nerves around the eye which supply the muscles that move the eye (the third nerve, the fourth nerve and the sixth cranial nerves). Damage to any of these nerves usually results in double vision. If the third nerve is affected there may also be a droopy eyelid (ptosis) and enlarged pupil on the affected side.

## Vasculitis and the Ear

Vasculitis can either affect the ear as part of a general illness, for example in Wegener's granulomatosis (now called granulomatosis with polyangiitis –GPA–), or can be a localised problem, for example autoimmune hearing loss. Problems with the ear can come before other features of disease in GPA and correct diagnosis is important in order to prevent hearing loss and more widespread illness. This article will discuss the different ways in which vasculitis can affect the ears and the different treatments available.

### Ear anatomy and function

It is worth starting by explaining the different names doctors give to parts of the ear (Figure 1) and the different types of hearing loss. The outer ear extends from the part that one can see up to the eardrum. The middle ear is the space between the eardrum and the skull, it contains the hearing bones or ossicles (malleus/mallet, incus/anvil and stapes/stirrup) and it is drained by the Eustachian tube. The inner ear forms part of the bone of the skull where the sound is converted into electric stimuli for transmission by nerves to the brain. Next to it are the semicircular canals and the rest of the balance organ.



**Figure 1. Anatomy of the ear (by M.Komorniczak published in Wikimedia commons)**

In terms of hearing loss it is divided into 'conductive loss', when the ear drum and/or the ossicles are affected; and 'sensorineural loss' when the cochlea, the nerve and/or the hearing centre in the brain are affected. In some cases the hearing loss is a mixture of conductive and sensorineural loss.

### How is vasculitis diagnosed in the ear?

Patients who present with vasculitis, which is limited to the ear, often have the same symptoms and show the same appearances as patients with infections and congestion in the middle ear. Very occasionally they have inflamed tissue behind the eardrum (Figure 2B). Another way vasculitis patients present is with sudden (over less than three days) nerve hearing loss.

The features that would point towards a diagnosis of vasculitis are an infection that does not respond to optimal treatment, greater pain than appearance of the ear would suggest and inflammatory tissue filling the middle ear. All patients that suffer from acute sensorineural hearing loss are treated with steroids. If the hearing responds well, an inflammatory process such as vasculitis is suspected, particularly if the hearing drops again on another occasion.

All patients suspected of GPA should have a urine dipstick done in the clinic and a chest x-ray as well as blood tests that include ANCA. However, in conditions such as Churg-Strauss syndrome (now called eosinophilic with polyangiitis –EGPA–) or limited GPA, these tests may not be helpful and biopsies may be required. The difficulty is that biopsies are often negative because vasculitis or granulomas may not be seen in the sample and what is seen could be caused by a number of inflammatory conditions. In these cases, close monitoring and repeat testing is required and occasionally treatment is started before having positive tests based on clinical findings.

### How do different forms of vasculitis affect the ear?

#### *Granulomatosis with polyangiitis (Wegener's granulomatosis)*

Acutely, patients may present with a painful discharging ear, infection or deafness. They often have nasal symptoms as well, that cause scarring of the Eustachian tube and congestion behind the eardrum. Some of these patients may have grommets (ventilation tubes through the ear drum) inserted that unfortunately, can result in constant ear discharge and little relief of deafness or discomfort.

During remission up to 60% of patients with GPA have involvement of the ears. Their complaints include persistent/recurrent discharge and hearing loss. The abnormalities seen when examining the eardrums are shown in Figure 2. The type and level of hearing loss seen in GPA is usually different in each ear and does not have a characteristic pattern seen in other conditions (for example with increasing age we tend to see patients with hearing loss that affects the same frequencies in both ears).



*Figure 2. Images of the tympanic membrane: A. congestion in the middle ear, B. Soft tissue mass in the middle ear, C. Chronically retracted tympanic membrane, D. Infected, retracted tympanic membrane with two small perforations inferiorly*

#### *Eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome)*

Ear disease in EGPA, can be also limited to the ears or part of a widespread disease. The symptoms are similar to those seen in GPA, but these patients seem to have a higher incidence of chronic inflammation in the middle ear and pain in the middle ear. The appearances and type of hearing loss is the same as that seen in GPA i.e. no characteristic pattern.

#### *Microscopic polyangiitis*

Patients with this type of ANCA vasculitis do not suffer from the destructive effects that GPA patients do. However, they may develop sensorineural hearing loss as a consequence of inflammation of the small vessels supplying the hearing nerve.

#### *Other symptoms*

There are other symptoms that can affect the ear, but they are considerably less common and are not associated with a particular type of vasculitis or in most cases with a vasculitic process. These include tinnitus (a sensation of sound in the ear without a stimulus) and vertigo. Tinnitus can affect anyone, but it is often associated with hearing loss. Vertigo takes many forms and the different inner ear pathologies present in the same way as in the general population. Rarely vasculitis can affect the nerve of the inner ear balance organ and cause similar symptoms to labyrinthitis (nausea and vomiting and a sensation of movement that lasts for a few days).

#### **How is it treated?**

Once the diagnosis is made, the initial treatment consists of systemic immunosuppression either with steroids alone or with immunosuppressants such as cyclophosphamide or methotrexate.

#### *Conservative*

Infections with ear discharge with or without a hole in the eardrum are treated antibiotic drops. If the infection is not settling or the canal is full of pussy discharge, the treatment involves cleaning the ear with suction in the ENT clinic and antibiotic creams or different antibiotic drops. If the infection is in the middle ear and does not settle on its own, it may require oral antibiotics. The treatment is expected to work within a week, but in the case of deep infections it may take a few weeks of intense treatment. However, one should consider low-grade activity if the infections do not settle as expected.

Hearing aids are useful in patients with conductive (middle ear problems) or sensorineural hearing loss ('nerve' hearing loss). Patients with constant ear discharge, who require aiding, may benefit from implantable devices. Hearing aids can also be helpful in patients with very intrusive tinnitus and if the patient has hearing loss, they are the first line of treatment. In patients whose tinnitus does not let them sleep or concentrate a referral to a specialist in tinnitus may be helpful. The specialists are usually audiologists who will try and find the trigger, provide information and sometimes white noise devices.

#### *Surgical*

Generally surgery is avoided in patients with vasculitis, but in patients who have been in remission for over 12 months and have a persistent discharge, surgery to repair the eardrum may be considered appropriate. In patients with congestion in the middle ear (glue ear) who are also in remission, a ventilation tube (grommet) may be helpful. There is not much evidence to inform the management decisions, so each individual case is considered with the advice of the medical team. In addition, patients may suffer from conditions affecting the rest of the population, such as, cholesteatoma (a condition where the ear drum retracts into the middle ear and traps skin within it). In these cases surgery should be considered with the advice from the medical team on medications before, during and after surgery.



### *Outcome*

Nerve related hearing loss can recover with corticosteroids in some cases, particularly when treated within a few days, but in most cases it does not recover completely. Conductive hearing loss usually implies damage to the eardrum or the ossicles or congestion behind the eardrum. If there is only glue ear with no other damage, grommets can potentially return the hearing to normal or to the level the nerve functions. If a more extensive procedure to repair the eardrum or the ossicles is required, the chances of improved hearing to normal levels are lower (between 50-80% will benefit five years after the surgery). The chance of repairing the eardrum successfully is quoted as 85% in the general population, but there are no published case series in vasculitis.

Regarding tinnitus, most the time the brain learns to ignore it and it becomes part of the background noise. In patients whose tinnitus is very intrusive the therapies mentioned above can aid the process of reducing the tinnitus to the background.

### **Is there anything I can do to protect my hearing?**

For patients who suffer from chronically discharging ears it is advisable to cover the opening of the ear canals with cotton wool smeared in Vaseline during showers or baths. If the patient is a keen swimmer personalised swimming moulds can be helpful. Cotton buds do not help to clean the ears and often cause more harm by traumatising the ear canal or pushing wax deep in the ear canal. No harm is done by using or not using hearing aids, but if people become withdrawn because they cannot follow a conversation, then this might be a good time to consider them.

*"Vasculitis and the Ear" has been written for Vasculitis UK by Dr Marcos Martinez Del Pero, Specialist Registrar in ENT, East of England Deanery, Cambridge, UK*

## Caring for a family member with vasculitis



Vasculitis does not just affect the patient. It has consequences for carers, families and also friends. Carers also face difficulties and problems dealing with the vasculitis patient. These vary depending on the stage at which the patient is at any particular time. The problems and difficulties encountered at first diagnosis change over time. Particularly at the diagnosis stage the carer often feels as bewildered as the patient. Below are some of the difficulties encountered by carers.

### **Initially before and during first diagnosis and initial treatment**

There are many problems for carers when coping with a very ill person who has no idea what is happening to them whilst the carer also has no idea what is happening, these include:

- Trying to take on board everything the medics are saying because the person with vasculitis is too ill to listen, talk, comprehend or is possibly unconscious
- Nursing a very ill person who is probably bed ridden or in hospital and the carer is probably trying to hold a job down and also taking care of the family at the same time
- Trying to explain to the immediate family what is happening when the carer doesn't have a clue themselves
- Coping with a very ill person who is in panic because they think they might die and the carer is also thinking the same, but explaining that everything will be ok and also trying to convince the family that the loved one won't die
- Helping the ill person to come to terms with the initial diagnosis and treatment whilst the carer is trying to come to terms with it themselves
- Coping with the big mood swings... denial.....anger....depression experienced by the vasculitis patient
- Ensuring that the ill person does not excluded the carer physically and emotionally.

### **After the initial diagnosis and treatment**

After initial diagnosis the carer and the one being cared for often face additional and on-going problems and difficulties. For the carer these can include:

- Helping the vasculitis patient to come to terms with taking masses of drugs whilst trying to come to terms with this situation themselves
- Trying not to worry about the future even though it is obvious that there may be difficulties, ie family, being able to work, paying bills, obtaining benefits etc
- Trying to be supportive with all hospital appointments and visits whilst possibly trying to hold down a job and juggle the family (school etc)
- Still coping with the denial.....anger.....depression.....sadness of the vasculitis patient
- Trying to convince the family that although the ill person may appear to be well they are not perhaps as well as they might appear
- Looking for support within the family
- Not knowing where to go for support and advice outside the family
- The carer wanting to talk to someone who knows how they feel

- Helping the vasculitis patient to cope with any eventual relapses/flare and never being sure if the problem is the vasculitis or something else causing the problem
- Worrying from day to day if the person who has vasculitis is ok and coping.

**Some organisations which offer carers support and help:**

- Crossroads and the NHS "caring for carers" gives advice, support and respite for all carers nationally. There are local offices throughout the UK
- Bernardo's gives advice support and respite for child carers

Useful links:

[www.Crossroads.org.uk](http://www.Crossroads.org.uk)  
[www.nhs.uk/carersdirect](http://www.nhs.uk/carersdirect)  
[www.barnardos.org.uk](http://www.barnardos.org.uk)

***Vasculitis UK offers one-to-one support for patients and carers. This support is given by a vasculitis patient or by the carer of a vasculitis patient. Contact John Mills (Chair, Vasculitis UK) for more information (see contacts page for details)***

## Children and young people with vasculitis



Vasculitis generally affects adults, but an increasing number of younger people and children are being diagnosed. This can have an impact on their schooling, general behaviour and future fertility. It is also a time when parents need to be aware of the services and benefits available. Government benefits have already been covered. However, specific to children and young people are the following:

### School support

It is essential to discuss the nature of the disease and the problems associated with the head and class teachers. This will enable a method of support to be developed, possible reduced hours, home tutoring or private tuition. Where

school support is not forthcoming or insufficient the Local Education Authority (LEA) should be contacted. The LEA has a Medical Referrals Team (MRA) who will liaise with the school. A letter from the consultant treating the child/young person will be necessary. Most LEAs provide some form of home tutoring but these vary from authority to authority.

### Special needs

It is essential to identify, assess and make provision for children/young persons requiring Special Educational Needs (SEN). There is an SEN Code of Practice to which schools, early education settings, LEAs and others must, by law, adhere to.

Details of all information on special needs are available from the LEA or the schools Special Education Needs Co-ordinator (SENCO) or Learning Support Co-ordinator. Web site information: [www.gov.uk/childrens-services/special-educational-needs](http://www.gov.uk/childrens-services/special-educational-needs)

[www.gov.uk/children-with-special-educational-needs/overview](http://www.gov.uk/children-with-special-educational-needs/overview)

In addition a podcast is available to listen to at: [www.cafamily.org.uk/?SEN\\_England.mp3](http://www.cafamily.org.uk/?SEN_England.mp3)

### Other services and allowances

The National Parent Partnership offers impartial advice, information and support to parents/carers with children with special needs: [www.parentpartnership.org.uk](http://www.parentpartnership.org.uk)

Education Maintenance Allowance may be available for 16-18 and some 19 year olds. Details from the school, college or training provider.

### Influenza vaccination for children

The nasal (Fluenz) vaccine is being recommended for children from 2 to 17 years old. However it should not be given to immunocompromised children. Nor should it be given to children who are in contact with immunocompromised adults since as it's live it may be transferred. Children can be given an injection of the inactivated vaccine as an alternative but it is known to be less effective for young people.

The NHS has produced a leaflet "Protecting Your Child Against Flu" - available from GP surgeries. This contains a section about children who shouldn't have the nasal vaccine.

### Transition: Getting it right for young people

*Improving the transition of young people with long term conditions from children's to adult health services*



### The aims of a transition programme

As they get older, children need to be involved increasingly in decisions about matters that affect them, so that by the time they are young adults they have learned to take responsibility for their own health. That task is more complicated for those with long term health problems and the price of getting it wrong can be much higher. An increasing number of children with complex disorders that only a few years ago were fatal in infancy or childhood now survive into adolescence and adult life, presenting new and unfamiliar challenges.

Young people move from a children's or young person's clinic, where they may have known their paediatrician and many other staff through much of their childhood and teens, to bigger clinics where they are surrounded by much older patients whose diseases may be very different from their own; they are less likely to see the same doctor at each visit; consultations may be shorter, and support and advice from staff may be less readily available. This is where the "transition programme" is helpful.

Transition can be defined as "a purposeful, planned process that addresses the medical, psychosocial and educational/vocational needs of adolescents and young adults with chronic physical and medical conditions as they move from child-centred to adult-oriented health care systems."

To view the full PDF transcript:

[webarchive.nationalarchives.gov.uk/20130107105354/http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH\\_4132145](http://webarchive.nationalarchives.gov.uk/20130107105354/http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_4132145)

#### **Further reading**

Wegener's granulomatosis in childhood - <http://onlinelibrary.wiley.com/doi/10.1002/art.24876/pdf>



Winston's Wish is the leading childhood bereavement charity and the largest provider of services to bereaved children, young people and their families in the UK.

They offer practical support and guidance to families, professionals and anyone concerned about a grieving child. They believe that the right support at the right time can enable young people to live with their grief and rebuild positive futures.

[www.winstonswish.org.uk/](http://www.winstonswish.org.uk/) Helpline: 08452 03 04 05 **Winston's Wish Head Office:** 4th Floor, St James's House, St James Square, Cheltenham, Gloucestershire, GL50 3PR. General Enquiries: 01242 515157 Fax: 01242 546187 Email: [info@winstonswish.org.uk](mailto:info@winstonswish.org.uk)

#### **Vasculitis UK - Leaflet for children with vasculitis**

The Trust has produced a leaflet especially for children diagnosed with vasculitis. Although it is worded for the child patient it may help to explain to children what is happening when a parent has been diagnosed with one of the vasculitides. The pamphlet is available from John Mills (Chair) - see contact details page.

## Health Service and other Organisational Support

### Prescription charges and exemptions

***Prescription charges and exemptions only apply in England. In Northern Ireland, Scotland and Wales there are no charges for prescriptions.***



In England free NHS prescriptions are available if, at the time the prescription is dispensed, you:

- are 60 or over
- are under 16
- are 16-18 and in full-time education
- are pregnant or have had a baby in the previous 12 months and have a valid maternity exemption certificate (MedEx)
- have a specified medical condition and have a valid medical exemption certificate (MedEx), eg (*note: some contain exceptions*)
- have a permanent fistula (eg, laryngostomy), Diabetes insipidus, Diabetes mellitus, Hypoparathyroidism, Myxodema, epilepsy requiring continuous anticonvulsive therapy
- have a continuing physical disability which means you cannot go out without help from another person and have a valid MedEx
- hold a valid war pension exemption certificate and the prescription is for the accepted disability, or the individual is an NHS inpatient.

You are also entitled to free prescriptions if you or your partner (including civil partners) are named on or are entitled to an NHS tax credit exemption certificate or a valid HC2 certificate (full help with health costs), or you receive either:

- Income Support
- Income-based Jobseeker's Allowance
- Income-related Employment and Support Allowance, or
- Pension Credit Guarantee Credit.

You are also issued with a MedEx if you are undergoing treatment for cancer. This includes treatment for the effects of cancer or for the effects of cancer treatments. Vasculitis is not a disease which automatically qualifies for free prescriptions.

Full details on NHS prescription charges and exemptions in England (including a translation link to several languages) can be found at:

[www.nhs.uk/nhsengland/Healthcosts/pages/Prescriptioncosts.aspx](http://www.nhs.uk/nhsengland/Healthcosts/pages/Prescriptioncosts.aspx)

Phone **0845 601 8076**

Information pamphlets are also available from pharmacists, local post offices, health authorities or benefits agencies.

### Pre-paid prescriptions

***Pre-paid prescriptions only apply in England. In Northern Ireland, Scotland and Wales there are no charges for prescriptions.***

If you do not qualify for free prescriptions and you are resident in England, it is possible to purchase a "pre-payment certificate (PPC)". A PPC could save money where more than four prescription items are required in three months, or fourteen or more items are required in twelve months. Leaflet HC12 gives full details including charges and can be obtained from Job Centre Plus offices, NHS hospitals, GP, dentist or optician.

PPCs are available by 10 monthly direct debit instalment payments. The prescription prepayment certificates allow an individual to obtain all the prescriptions they need for £2 per week.

Full details, including current charges (including a translation link to several languages) can be found at:

[www.nhs.uk/NHSEngland/Healthcosts/Pages/Abouthealthcosts.aspx](http://www.nhs.uk/NHSEngland/Healthcosts/Pages/Abouthealthcosts.aspx)

Phone **0845 850 0030**

## Community Nurse



If nursing care or support at home is required, a community nurse could help.

Community nurses are registered nurses who work in the community: in people's homes, in schools and in local surgeries and health centres. The people they work with may be ill or disabled. Community nurses also look after people whose health may be particularly vulnerable, such as older people, children, or people with learning disabilities. They visit people at home to provide health care, for example, changing dressings or giving injections and help people get any home nursing aids and equipment they need.

Community nurses can provide help and advice on a wide range of health issues. They may also teach families and carers basic care giving skills. They work closely with GPs, local social services and hospitals. The GP can refer the patient to a community nursing service.

## Hospital Transport

There is a scheme which offers financial assistance to patients in order to help them travel to hospital, but the need for that transport will be assessed. For patients where there are medical needs for ambulance transport the particular Trust concerned is responsible.

Help is available under the Hospital Travel Costs Scheme (HTCS), to those patients who do not have a medical need for ambulance transport and for those who cannot meet the cost of travel to hospital.

Form HC11 - Help with Health costs should be available at the GP's surgery. It does not just cover the cost of travel – it also covers other aspects of health needs.

Further details are available at:

[www.nhsbsa.nhs.uk/HealthCosts/1558.aspx](http://www.nhsbsa.nhs.uk/HealthCosts/1558.aspx)

Patients who are eligible would be those in receipt of:-

- Income support
- Income based jobseekers allowance
- Tax credit – working tax credit or child tax credit
- Pension credit
- Guarantee credit

Other patients who are on a low income may be entitled to partial or full reimbursement.

## Consultations outside NHS Area

In England, if you are travelling outside your NHS area for a consultation, e.g. an appointment at Addenbrooke's or Birmingham, you may be able to claim travel and possibly overnight accommodation expenses.

For full details visit: [www.nhs.uk/NHSEngland/Healthcosts/Pages/Travelcosts.aspx](http://www.nhs.uk/NHSEngland/Healthcosts/Pages/Travelcosts.aspx)

Some hospitals do have special overnight accommodation and also reduced parking fees for patients. For example there is accommodation within the grounds of Addenbrooke's for patients who have to travel some distance for their appointment and require an overnight stay: "Pemberton House - Frank Lee Leisure and Fitness"

[www.cuh.org.uk/corporate-information/services/non-clinical-services/accommodation/visitor-accommodation](http://www.cuh.org.uk/corporate-information/services/non-clinical-services/accommodation/visitor-accommodation)

In addition on production of an appointment letter, you can receive a special parking ticket (maximum cost of around £3.50 irrespective of the length of the stay). A visitor to the Addenbrooke's car park (run by NCP) can pay up to £9 a day!

At other hospitals details should be available at reception or contact Hospital Administration to ascertain if similar facilities are available.

***In Northern Ireland, Scotland and Wales you should contact the Patient Expenses Officer (Finance Department) at your hospital.***

## Equipment and Services

The Red Cross volunteer-led medical equipment service provides wheelchair hire and short-term loans of equipment in almost 1000 outlets in the UK, helping tens of thousands of people every year.

The medical equipment service helps people return to their own homes after illness or surgery, enables them to go on holiday with friends or family and promotes independence. The main types of equipment provided include:

- wheelchairs
- backrests
- bath seats
- walking sticks and frames
- commodes, bedpans and urinals.

For further details visit: [www.redcross.org.uk](http://www.redcross.org.uk) Or write to: British Red Cross, UK Office, 44 Moorfields, London EC2Y 9AL

Tel: 0844 871 11 11 (+ 44 2071 3879 00 from abroad)  
Fax: 020 7562 2000 Minicom: 020 7562 2050



## Workplace issues

How much an employee tells the employer about their illness is a personal choice. However, if the employer knows the difficulties being encountered the better able he/she will be to offer help and advice regarding the employee's ability to remain in employment or to arrange more suitable duties. If a company has an occupational health physician it may be helpful to speak to him/her if encountering difficulties undertaking normal duties.

Further advice can be obtained from your local Citizens Advice Bureau.



## Statutory Sick Pay

If you are unable to work because of illness it may be possible to get Statutory Sick Pay (SSP). It is paid by the employer and can be paid for up to 28 weeks.

If you are working under a contract of service (even if only for a short period and have done some work), you are entitled to SSP if the following apply:

- You have been sick for at least four days in a row (including weekends and bank holidays and days that they do not normally work)
- You have average weekly earnings of at least £97 a week (as at 2011)

Average weekly earnings are calculated by using earnings in the eight weeks before sickness began.

To get SSP you must:

- tell the employer that you are sick
- if asked by the employer you must provide some form of medical evidence from the eighth day of illness.

## Occupational sick pay schemes

If the employer has a sick pay scheme, which is equal to, or more than SSP, they do not have to operate the SSP scheme. The employer may also have different rules for payment, which the employee must keep to receive payment.

If you are sick after 28 weeks of occupational sick pay, or if this ends earlier and you are not entitled to SSP, the employer must provide a form (SSP1) which enables you to claim Employment and Support Allowance.

If working abroad, you may be able to get SSP if the employer pays National Insurance contributions on your behalf. It is possible to go abroad to visit and still claim SSP provided proof of continued sickness is provided. Serving members of the Armed Forces cannot get SSP, but members of their families may qualify, if they satisfy the conditions for payment.

Full details on SSP are available in English and Welsh at: <https://www.gov.uk/statutory-sick-pay>

## Government Benefits

***Vasculitis UK can provide, to its Members, free information from an independent organisation "Benefit and Work". The documentation gives help on claiming benefits and appeals procedures for DLA. Please contact Hon Secretary - Jann Landles (see contact details).***

## **Disability Living Allowance (DLA)**

Disability Living Allowance is a tax-free benefit for disabled children and adults who need someone to help look after them, or have walking difficulties.

Since April 2013 Disability Living Allowance is only payable to children under 16 and adults aged 65 and over who were in receipt of DLA prior to April 2013. All other claimants are now being assessed for Personal Independence Payments (PIPs)

Disability Living Allowance is a tax-free benefit for disabled children and adults who need someone to help look after them, or have walking difficulties.

You may get DLA if:

- you have a physical disability (including a sensory disability, such as blindness) or mental disability (including learning disabilities), or both
- the disability is severe enough for you to need help caring for yourself or someone to supervise you, for your own or someone else's safety, or you have walking difficulties, or both
- you are under 16 and over 65 when claiming.



Normally, you must have had these care or supervision needs or walking difficulties for at least three months and you are likely to continue for at least a further six months. However, in the case of terminal illness, there are special rules for claiming the benefit.

If you are aged 65 or over, you may be able to get Attendance Allowance.

Complete details, including rates payable and how to claim can be viewed on the "Directgov" website: [www.gov.uk/](http://www.gov.uk/)

Alternatively a "claims pack" can be obtained by phoning the Benefits Enquiry Line. They are open Monday to Friday 8.30am to 6.30pm and Saturdays 9.00am to 1.00pm: Telephone: 0800 88 22 00 Textphone: 0800 24 33 55

When applying for disability benefits it is advisable to get help from a local disability group, Citizens Advice Bureau or other similar organisations. The forms are complicated and difficult to complete. The problems encountered by vasculitis patients when claiming DLA cannot be over-emphasised. It is suggested that the patient obtains a report from the medical consultant outlining current difficulties and long term prognosis. Vasculitis patients often look well but have significant current and long term limitations.

These benefits are applicable throughout the UK. Although further details for Northern Ireland can be found at: [www.nidirect.gov.uk/](http://www.nidirect.gov.uk/)

**From April 2013 DLA was replaced by Personal Independence Payments (PIPs) - see PIP section.**

### **Motability**

Persons receiving the higher rate of mobility allowance can purchase a brand new car without the worry of servicing, tax, insurance etc. Purchase is via an accredited dealership and once approved the vehicle is paid for using the mobility allowance. Depending on the cost of the vehicle additional contributions may be necessary. Details from: [www.motability.co.uk](http://www.motability.co.uk) or phone 01279 635666

## Road Fund licence

If you are in receipt of:

- higher rate of the mobility component of Disability Living Allowance or
- War Pensioners Mobility Supplement

it is possible to obtain an exemption certificate whereby you are able to tax your own vehicle for free.

Full details can be found at:

[www.direct.gov.uk/en/Motoring/OwningAVehicle/HowToTaxYourVehicle/DG\\_4022121](http://www.direct.gov.uk/en/Motoring/OwningAVehicle/HowToTaxYourVehicle/DG_4022121)

## The Blue Badge Scheme

The Blue Badge Scheme is an important service for people with severe mobility problems that enables badge holders to park close to where they need to go. The scheme operates throughout the UK and is administered by local authorities who deal with applications and issue badges.

### About the Blue Badge scheme

The Blue Badge Scheme only applies to on-street parking.



Badge holders may park on single or double yellow lines for up to three hours in England and Wales, except where there is a ban on loading or unloading. There is no time limit for parking on yellow lines in Scotland. Where a time limit is in force, both the Blue Badge and the parking clock, set to show the arrival time, must be displayed.

Badge holders may park for free and for as long as they need to at on-street parking meters and pay-and-display machines.

While the scheme operates throughout the UK, there are small variations in its application in England, Wales, Scotland and Northern Ireland. Please see the relevant website for further information. The information on this page relates to England unless specified.

Full details are available from:

[www.direct.gov.uk/en/AdvancedSearch/Searchresults/index.htm?fullText=disabled+people+motoring+and+transport](http://www.direct.gov.uk/en/AdvancedSearch/Searchresults/index.htm?fullText=disabled+people+motoring+and+transport) or local council offices.

## Attendance Allowance

Attendance Allowance is a tax-free benefit. You may get Attendance Allowance if you're aged 65 or over and need help with personal care because you're physically or mentally disabled.

### Who can get Attendance Allowance?

You may get Attendance Allowance (AA) if the following apply:

- you have a physical disability (including sensory disability, such as blindness), a mental disability (including learning difficulties), or both
- your disability is severe enough for you to need help caring for yourself or someone to supervise you, for your own or someone else's safety
- you are aged 65 or over when you claim

Attendance Allowance isn't usually affected by any savings or income you may have.

If you're under age 65, you may be entitled to other benefits.

Full details are available at the UK government website: [www.gov.uk/browse/benefits/disability](http://www.gov.uk/browse/benefits/disability) scroll down the list for Attendance Allowance.

**Changes to Benefits as from April 2013** - During April 2013 many government benefits were changed. Further details on government benefits can be found at Citizens Advice - [www.adviceguide.org.uk/england/benefits\\_e/benefits\\_sick\\_or\\_disabled\\_people\\_and\\_carers\\_ew/benefits\\_for\\_people\\_who\\_are\\_sick\\_or\\_disabled.htm](http://www.adviceguide.org.uk/england/benefits_e/benefits_sick_or_disabled_people_and_carers_ew/benefits_for_people_who_are_sick_or_disabled.htm)

When visiting the Citizens Advice website ensure you select the UK country in which you are resident.

## Disabled Students Allowances (DSAs) in higher education

Disability Living Allowance is a tax-free benefit for disabled children and adults who need someone to help look after them, or have walking difficulties.

Since April 2013 Disability Living Allowance is only payable to children under 16 and adults aged 65 and over who were in receipt of DLA prior to April 2013. All other claimants are now being assessed for Personal Independence Payments (PIPs) - [www.vasculitis.org.uk/living-with-vasculitis/government-benefits-personal-independence-payments](http://www.vasculitis.org.uk/living-with-vasculitis/government-benefits-personal-independence-payments)

Or visit HM Government website: [www.gov.uk/browse/benefits/disability](http://www.gov.uk/browse/benefits/disability)

## Employment and Support Allowance



Employment and Support Allowance (ESA) provides financial help to people who are unable to work because of illness or disability.

It also provides personalised support to those who are able to work. Details are given below, but more information about ESA can be found on the Government's website:

[www.gov.uk/browse/benefits/disability](http://www.gov.uk/browse/benefits/disability)

### About ESA

ESA offers you a personalised support and financial help, so that you can do appropriate work, if you are able to. It gives you access to a specially trained personal adviser and a wide range of further services including employment, training and support.

ESA involves a medical assessment called the Work Capability Assessment which assesses what you can do, rather than what you cannot, and identifies the health-related support needed. Most people claiming ESA will be expected to take steps to prepare for work, including attending work-focused interviews with a personal adviser.

Under ESA where the claimant has an illness or disability that severely affects their ability to work, they will get increased financial support. The claimant will not be expected to prepare for a return to work. They can volunteer to do so at any point if they wish to do so.

### Who can get ESA?

You may be able to get ESA if you have an illness or disability that affects your ability to work. You may be able to claim ESA if any of the following apply to you:

- Statutory Sick Pay has ended, or you cannot get it
- are self employed or unemployed
- have been getting Statutory Maternity Pay (SMP) and have not returned to work because you have an illness or disability which affects your ability to work
- are under State Pension age

You must also either:

- have had an illness or disability which affects your ability to work for at least four consecutive days (including weekends and public holidays)
- be unable to work for two or more days out of seven consecutive days
- be getting special medical treatment

If you are aged between 16 and 20 (or under 25 if you were in education or training at least three months immediately before turning 20), you must:

- have been too ill to work because of an illness or disability for at least 28 weeks (this limitation only applies to contribution-based ESA, but may still be eligible for income-based ESA)
- have been too ill to work before they turned 20 (or 25 if you were in education or training at least three months immediately before turning 20)

### Entitlement conditions

There are two types of ESA:

**Contribution-based ESA** You may be entitled to claim contribution-based ESA if you have paid sufficient National Insurance contributions.

**Changes to Contribution-based Employment and Support Allowance** The government limited the period for which Contribution-based ESA can be paid in some circumstances. These changes were part of the Welfare Reform Act, enacted in March 2012. The changes are:

- to limit the amount of time people in the Work-Related Activity Group (ie those not in the Support Group) can receive Contribution-based ESA to 365 days, and
- to remove the special contribution condition and prevent any new claims for ESA on grounds of youth.

For full details on the changes to Contribution-based ESA see: UK government disability website: [www.gov.uk/browse/benefits/disability](http://www.gov.uk/browse/benefits/disability)

**Income-based ESA** You may be entitled to claim income-based ESA if you are receiving insufficient money, or you have not paid sufficient National Insurance contributions and you satisfy the entitlement conditions. This means that you have savings of less than £16,000 and if you have a partner or civil partner, they work for less than 24 hours a week on average.

#### **ESA rates and how to claim**

The amount paid depends on individual circumstances. It also depends on what effect the disability has on the claimant's ability to do any work.

Telephone: 0800 055 6688 Textphone: 0800 023 4888 Claims can be made in Welsh. Telephone: 0800 012 1888

These benefits are applicable throughout the UK. However, further details for Northern Ireland can be found at: NI Direct - [www.nidirect.gov.uk/](http://www.nidirect.gov.uk/)

Indepth information about ESAs can be found at: Citizens Advice Bureau: [www.adviceguide.org.uk/england/benefits\\_e/benefits\\_sick\\_or\\_disabled\\_people\\_and\\_carers\\_ew/benefits\\_for\\_people\\_who\\_are\\_sick\\_or\\_disabled.htm](http://www.adviceguide.org.uk/england/benefits_e/benefits_sick_or_disabled_people_and_carers_ew/benefits_for_people_who_are_sick_or_disabled.htm)



**Disability Alliance:** "Disability Alliance" provides information on social security benefits, tax credits and social care to disabled people, their families, carers and professional advisers. For help or information about this organisation go to:

[www.disabilityalliance.org/benefits.htm](http://www.disabilityalliance.org/benefits.htm)

## Personal Independence Payments (PIPs)



The government is introducing Personal Independence Payment to replace Disability Living Allowance.

Disability Living Allowance will end for everyone of working age even if they have an indefinite period award.

Working age means everyone who is aged 16 to 64 on the day Personal Independence Payment is introduced.

There are no current plans to replace Disability Living Allowance for children under 16 and people aged 65 and over who are already receiving Disability Living Allowance.

Personal Independence Payment is based on an assessment of individual need. The new assessment will focus on an individual's ability to carry out a range of key activities necessary to everyday life. Information will be gathered from the individual, as well as healthcare and other professionals who work with and support them. Most people will also be asked to a face to face consultation with a trained independent assessor as part of the claim process.

### Introduction of PIP

#### For new claims

From April 2013 - Personal Independence Payment were introduced for new claims in Merseyside, North West England, Cumbria, Cheshire and North East England. During this period new claimants in all other parts of the country continued to claim Disability Living Allowance.

From June 2013 - new claims for Personal Independence Payment were taken from all parts of the country.

#### For existing DLA claimants

Existing Disability Living Allowance claimants who are aged 16 to 64 on 8 April 2013 were affected by the introduction of Personal Independence Payment, even if they had an indefinite or lifetime award of DLA.

Personal Independence Payment is being introduced in stages over a number of years and claimants will be contacted with more information.

From October 2013 - if there is a change in how a health condition or disability affects an individual, or they reach the end of an existing award of DLA, then they will be invited to claim Personal Independence Payment.

From 2015 - The Benefits Agency will start to contact everyone else receiving DLA (unless they report a change in how their health condition or disability affects them, or if their award is due to end). They will write to individuals in plenty of time and you do not need to contact DWP now.

There is no automatic transfer from Disability Living Allowance to Personal Independence Payment.

For full information on the introduction and background to PIPs see: UK government website - [www.gov.uk/browse/benefits/disability](http://www.gov.uk/browse/benefits/disability)

In depth information about PIPs is available on the Citizens Advice Bureau website:

[www.adviceguide.org.uk/england/benefits\\_e/benefits\\_sick\\_or\\_disabled\\_people\\_and\\_carers\\_ew/benefits\\_for\\_people\\_who\\_are\\_sick\\_or\\_disabled.htm](http://www.adviceguide.org.uk/england/benefits_e/benefits_sick_or_disabled_people_and_carers_ew/benefits_for_people_who_are_sick_or_disabled.htm)

When visiting the CAB website ensure you select the UK country in which you are resident.

## Carers Allowance

From April 2010 the Carer's Allowance was extended to cover more carers. Carer's Allowance is a taxable benefit to help people who look after someone who is disabled. The claimant does not have to be related to, or live with, the person being cared for.

A carer may be able to get Carer's Allowance if they are aged 16 or over and spend at least 35 hours a week caring for a person who gets either:

- Attendance Allowance
- Disability Living Allowance at the middle or highest rate for care
- Constant Attendance Allowance at or above the normal maximum rate with an Industrial Injuries Disablement Benefit
- Constant Attendance Allowance at the basic (full day) rate with a War Disablement Pension



Carer's Allowance is not available if the individual is in full-time education with 21 hours or more a week of supervised study or earn more than £100 a week after certain deductions have been made - for example Income Tax.

This benefit is applicable to the whole of the UK.

Further details, including the amount payable can be found at:

[www.gov.uk/browse/benefits/disability](http://www.gov.uk/browse/benefits/disability)

## Social Care and Support

Changes have recently come into force regarding care available from councils. The new system involves direct payments i.e. personally arranging for own care and services

Direct payments are made by councils to people receiving social care services, instead of the council providing the service directly. These are local council payments for people who have been assessed as needing help from social services and who would like to arrange and pay for their own care and support services instead of receiving them directly from the local council.

A person must be able to give their consent to receiving direct payments and be able to manage them even if they need help to do this on a day-to-day basis.

If a person already receives social services the local council must offer the option of direct payments in place of the services currently received. There are some limited circumstances where this choice is not given. The council concerned will be able to give details where these apply.

*Where the individual is not receiving social services they should contact their local council requesting that their needs be assessed. Social services - and therefore direct payments - are normally available if the individual is:*

- disabled and aged 16 or over
- a carer aged 16 or over, including people with parental responsibility for a disabled child
- an older person



If the local council has decided that a claimant does not need social care services, it will not offer direct payments. If an individual considers that their needs or circumstances have now changed, they should ask the council for a new assessment.

The amount received will depend on the outcome of the assessment the council makes of the individual's needs.

If applying for services for the first time, the social worker should discuss the direct payments option with the individual when they assess the care needs.

Information for local council websites in England is given in the link below. Further information is also available together with how to apply online:

[www.local.direct.gov.uk/LDGRedirect/index.jsp?LGSL=287&LGIL=0&Samp:erviceName=Apply+for+direct+payments](http://www.local.direct.gov.uk/LDGRedirect/index.jsp?LGSL=287&LGIL=0&Samp:erviceName=Apply+for+direct+payments)

The money is for the individual to use to pay for the services and equipment which will meet the needs the local council has assessed as being appropriate.

As a general principle councils should let the individual choose how best to meet their assessed needs as long as they are satisfied that agreed support arrangements are being met.

Direct payments cannot be used to pay for permanent residential accommodation. Direct payments may be used to pay for occasional short periods in residential accommodation, if the council agrees that is what is needed.

Unless the council decides that exceptional circumstances make it necessary, direct payments cannot be used to pay for a service from:

- a spouse (husband or wife)
- a civil partner
- a partner with whom the claimant lives as a couple
- a close relative with whom the claimant lives, or the spouse or partner of that close relative

Where direct payments are received, the recipient will need to account for the money they spend. The council will explain what records need to be kept and what information they expect. Examples include timesheets signed by personal assistants, or receipts for services from agencies.

The council has to be satisfied that the needs for which the direct payment is given are being met. They should state how they will achieve this. This may involve a visit to the home of the individual.

Carers aged 16 or over may be eligible for direct payments for themselves. Direct payments cannot be used to buy services for the person being cared for. They can only be spent on getting the support the actual carer has been assessed as needing.

Direct payments are not a replacement of income and therefore do not affect any other benefits being received.

For further information on Social Care in Northern Ireland see [www.hscni.net/](http://www.hscni.net/)

For Scotland see [www.scotland.gov.uk/Topics/Health/Support-Social-Care](http://www.scotland.gov.uk/Topics/Health/Support-Social-Care)

For Wales see [wales.gov.uk/?lang=en](http://wales.gov.uk/?lang=en)

### **Other Benefits and Allowances**

There are other benefits and allowances available. What can be claimed will depend on individual circumstances. The Citizens Advice Bureau can help regarding the benefits available and they help the claimants to complete the forms correctly.

A complete list of benefits which are available in the UK can be found at the following sites:

[www.gov.uk/browse/benefits/disability](http://www.gov.uk/browse/benefits/disability)

## Insurance

It can be difficult to obtain travel, life and health insurance when suffering from diseases such as vasculitis. Often pre-existing conditions are excluded and, if covered, the additional supplements can be high. However, the cost of medical treatment, when travelling abroad can cost the patient requiring medical treatment, many thousands of pounds.

### Travel insurance

There are a number of companies specialising in travel insurance for patients with pre-existing medical conditions. The following companies have been used by Vasculitis UK members and we have received good reports on their services and conditions. Patients are advised to shop around for the best deals.

**Just Travel Insurance:** Just Travel Ltd. Victoria House, Sunderland, SR1 2QF  
phone: 0800 231 5535 [www.conditionscovered.co.uk/](http://www.conditionscovered.co.uk/) *This company will donate a small percentage to Vasculitis UK for Members mentioning Vasculitis UK when taking out the policy.*

**Virgin Insurance:** Cover for pre-existing conditions and for over 65's. phone: 0844 888 3900  
<http://uk.virginmoney.com/virgin/travel-insurance/>

**The Post Office** offers different levels of travel insurance. Like most insurers they have specific rules regarding pre-existing medical conditions which relate to the proposer and to any other member of the party to be travelling. Full details and application forms are available from the Post Office.

Many bank accounts offer special packages and often these include free holiday travel. Where pre-existing medical conditions exist many of these packages will carry a supplement payable to the actual company offering the insurance on behalf of the bank.

As with insurance providers, it is essential that the information given is absolutely correct. There have been instances where insurers have refused to pay because the illness or problem has not been fully disclosed. Where pre-existing conditions have been reported to the insurer it is best to request a written acknowledgement.

### Health insurance

Companies offering health insurance, such as BUPA, each have their own rules regarding acceptance of persons with pre-existing medical conditions. Each company will differ and the proposer should contact the company with which they are seeking cover.

### Mortgage protection/Loan protection/Occupational pension plans

As with all types of insurance, any pre-existing or previous illnesses will have to be declared. This may result in additional premiums being requested or claims involving these illnesses being excluded from the cover. Where long term insurance/protection is required the company may limit the cover available. Discussion with the representative of the company concerned is advised.



## Genetic Alliance UK "Asking the relevant questions, Insurance Project Report, 2010"

Genetic Alliance UK has undertaken some work on insurance and details can be found at:  
[www.geneticalliance.org.uk/docs/askingrelevantquestions.pdf](http://www.geneticalliance.org.uk/docs/askingrelevantquestions.pdf)

## Frequently asked questions (FAQs)

### What is vasculitis?

There are a number of vasculitis diseases (the vasculitides). All are characterised by inflammation of the blood vessels in one or more organ(s). The vasculitides are diseases involving the immune system whereby the immune system no longer provides defence against infection or other triggers and, therefore, the immune system fights itself.

### What is inflammation?

The process used by the body to protect against or eliminate foreign bodies, bacteria or viruses. The usual signs of inflammation include redness of the skin, swelling, feeling of warmth or pain in the affected areas.

### What are the different types of vasculitis?

The vasculitides can affect small, medium or large arteries. The main diseases are:

#### Large sized arteries

- Giant Cell Arteritis/Temporal Arteritis
- Polymyalgia Rheumatica
- Takayasu Arteritis

#### Middle sized arteries

- Kawasaki Disease
- Polyarteritis Nodosa

#### Small sized arteries

- Eosinophilic Granulomatosis with Polyangiitis (previously known as Churg-Strauss Syndrome)
- Microscopic Polyangiitis
- Granulomatosis with Polyangiitis (previously known as Wegener's Granulomatosis)
- Henoch-Schönlein Purpura

#### Less common vasculitides

- Anti GBM (Goodpastures Disease)
- Behçet's Syndrome
- Buerger's Disease
- Central Nervous System Vasculitis
- Cogan's Syndrome
- Cryoglobulinemia and Cryoglobulinaemic Vasculitis
- Urticarial Vasculitis

### What are the symptoms of vasculitis?

As there are a number of different vasculitis diseases and as they can attack most organs of the body the symptoms differ from vasculitis to vasculitis and from patient to patient. Some general symptoms include: tiredness, weakness, loss of appetite, weight loss and fever

### What drugs are used to treat vasculitis?

There are two stages in the treatment of vasculitic disease. The first stage is controlling the disease process and the second is maintaining remission and preventing relapse. Depending on the disease, the patient and the severity of the disease the initial stage drug regime can include low dose chemotherapy (cyclophosphamide), co-corticosteroids (prednisolone), methotrexate or Azathioprine. For maintenance of remission and to prevent relapse the drug regime includes methotrexate, azathioprine, cellcept (mycophenolate mofetil) and low dose co-corticosteroids (prednisolone) (see individual disease sections). Other drugs are used where patients are intolerant to the usual drugs or where these drugs have not been successful in controlling the inflammation, e.g. Rituximab.



**What causes vasculitis?**

In most cases of vasculitis the exact cause is not known. However recent research suggests that in most cases of vasculitis a genetic or environmental trigger (or a combination of both) may be the cause.

**How common is vasculitis?**

The vasculitides are rare diseases, with 10-15 new cases a year (per million population) being diagnosed with ANCA associated vasculitis.

**Who are affected?**

Some of the vasculitides affect males and females in equal numbers, whilst others tend to affect a higher proportion of males. The usual onset for vasculitis diseases generally is from age 50 onwards, but young people and infants can be affected.

**What is ANCA?**

Anti-neutrophil-cytoplasmic antibody. In ANCA associated vasculitis the B-cells produce antibodies that are directed against proteins found on the surface of neutrophils (a type of white blood cell).

**Does a negative ANCA mean I don't have vasculitis?**

No, some patients can be ANCA positive and others ANCA negative. Individual patients may be positive at one blood test and negative at another. The physician uses the ANCA test as a guide to treatment, along with other test results and by examination and discussion with the patient.

**Should the vasculitis patient avoid taking other drugs**

Drugs such as "over the counter" drugs should not be taken unless this has been discussed with the doctor or pharmacist.

**Is vasculitis hereditary?**

There is a possibility of a genetic link which might predispose individuals to developing one of the vasculitic diseases, but it is not considered that genetic predisposition alone is a factor.

**Why is vasculitis so difficult to diagnose?**

There are a number of vasculitic diseases and each can involve any number of organs in the body. In addition many of the symptoms the patient presents with are similar to many other illnesses or diseases. Vasculitis can mimic other diseases. As the treatment for vasculitis is by prescribing toxic drugs there are many diagnostic tests to be undertaken to rule out other diseases and to confirm the presence of vasculitis.

**Is vasculitis infectious or contagious?**

No

**Is there a cure for vasculitis?**

Although some of the vasculitides are self-limiting the majority cannot be cured. Research continues into the causes of vasculitis and treatments. Vasculitis is controlled by maintenance therapy.

**How often should I see my consultant?**

In the initial stages of treatment patients are seen at regular intervals by their consultants. When remission is achieved the period between consultations is increased, but even where patients are drug free they should still be monitored regularly.

**Which consultants treat vasculitis?**

This depends on the organs involved, but the main specialties include: nephrologist (renal), rheumatologist, ear, nose and throat, paediatrician, ophthalmologist, neurologist and respiratory physicians.

**Do consultants specialise in vasculitis?**

As the vasculitic diseases are rare most patients are treated by consultants whose main specialties include: nephrologist (renal), rheumatologist, ear, nose and throat, paediatrician, ophthalmologist, neurologist and respiratory physicians. However, a number of these consultants also specialise in treating patients with vasculitic diseases.

**Where will I receive my treatment?**

Usually patients are seen at local or area hospitals. Some patients are seen at national centres dealing with vasculitis. Where the patient is treated should be discussed between the patient and the GP.

**Are there centres which specialise in treating vasculitis?**

There are a number of national centres which specialise in treating vasculitis. Referral to these centres should be discussed with the GP.

### **Is research being undertaken into vasculitis?**

Yes. A considerable amount of research has been undertaken nationally and internationally into the causes, treatment and effects of vasculitis. National and international research projects are currently ongoing.

### **How often should I have blood tests?**

In the initial stages bloods monitoring is undertaken at regular intervals depending on the drug regime prescribed. The periodicity of blood testing when the patient is in remission depends on the patient, the maintenance therapy prescribed and other medical factors.

### **How do I interpret the results of blood tests?**

Your physician will interpret the results and discuss these with you. There are no absolute levels for the various tests but these can vary between patients and the laboratory doing the analysis. It is the change found from test to test in the individual which is important.

### **What tests/procedures are undertaken?**

These depend on the vasculitis and the symptoms.

### **What is a biopsy?**

This is a procedure whereby a small piece of tissue is taken from the affected area or organ, e.g. the skin or the kidney. The tissue is then examined under a microscope. A biopsy is the most conclusive way of ascertaining whether vasculitis is present.

### **What is a flare?**

A flare is where the disease has been relatively stable on maintenance therapy but there is a sudden change in the original symptoms or new symptoms are reported.

### **What is remission?**

After the initial treatment period, when the physician considers the disease is controlled, he/she may consider the patient to be in remission or inactive.

### **Can vasculitis be fatal?**

In some cases of severe disease if not diagnosed early and not treated correctly. With early diagnosis and appropriate treatment vasculitis is now rarely fatal. Many milder cases may cause damage to organs or discomfort but are not life-threatening.

### **Is vasculitis linked to cancer?**

No. However, there is a link between the use of chemotherapy and the onset of bladder cancer in later life, but modern dosages of chemotherapy have reduced this possibility considerably.

### **Is vasculitis like AIDS?**

No. Vasculitis involves the immune system being over-active. AIDS is an acquired disease where the immune system is under-active. AIDS can also be transferred to others whereas vasculitis cannot.

### **Can the vasculitis patient have another autoimmune disease?**

Yes.

### **Should I have vaccinations?**

Patients taking immunosuppressants **should NOT receive LIVE vaccines**. However, flu and pneumonia vaccines **are** recommended for vasculitis patients. Where the patient is in doubt about a particular vaccine this should be discussed with the GP.

### **Can vasculitis affect fertility?**

Taking cyclophosphamide and similar drugs may render the patient less fertile or infertile.

### **Is it advisable to use contraception during treatment?**

Whilst taking drugs such as cyclophosphamide it is essential to use effective contraception as the drugs will cause damage to the unborn child.

### **Can vasculitis affect the eyes?**

Yes some patients can have problems with their eyes.

### **Can vasculitis affect the hearing?**

Yes some patients can have problems with their hearing.

**Can children be diagnosed with vasculitis?**

Unfortunately a number of children are diagnosed with vasculitis each year.

**Is it possible to manage the pain of vasculitis?**

There are a number of ways to help manage pain.

**Is it possible to manage low mood and depression caused by vasculitis?**

Many vasculitis patients suffer from low mood or depression. This is a normal phenomenon and is often transient..

**What about oral health for the vasculitis patient?**

It is important to ensure good oral hygiene for all vasculitis patients. This is especially the case where there are pre-existing dental problems which may get worse due to steroids or immuno-suppressants.

**What about diet?**

Some patients may be prescribed a special diet and it is essential to keep to this eating regime. Diet-controlled food intake is important since too much will cause weight gain in addition to that experienced by the use of steroids. For further information.

**What about alcohol?**

Alcohol should not be taken with some of the drugs used to treat vasculitis. Where alcohol consumption is permitted this should be in moderation.

**Is there an increased chance of becoming diabetic?**

Some patients may become diabetic and this will be controlled by drugs and a healthy diet.

**Should I exercise?**

The vasculitis patient can easily become fatigued, but light exercise, walking, swimming etc will ensure your muscles stay strong and flexible. Any exercise should be within your capabilities.

**Should I have a DEXA (bone) scan?**

Patients taking high doses of steroids are at risk of developing osteoporosis. Therefore, periodic bone scans are recommended. The risk of developing osteoporosis and the best way to treat or monitor this should be discussed with the medical team.

## Website links

### National and International Information

- **Vasculitis UK** - [www.vasculitis.org.uk/](http://www.vasculitis.org.uk/)
- **RareRenal.org - Vasculitis Rare Disease Group** - [www.rarerenal.org/rare-disease-groups/vasculitis-rdg/](http://www.rarerenal.org/rare-disease-groups/vasculitis-rdg/)
- **Trinity Health Kidney Centre** - (Trinity College Dublin) - [www.medicine.tcd.ie/thkc/research/](http://www.medicine.tcd.ie/thkc/research/)
- **Vasculitis Awareness Ireland** - [www.vasculitis-ia.org/](http://www.vasculitis-ia.org/)
- **Arthritis Research UK** - [www.arthritisresearchuk.org/](http://www.arthritisresearchuk.org/)
- **Cortisone Information** - [www.cortisone-info.com/](http://www.cortisone-info.com/)
- **Vasculitis Foundation (USA)** - [www.vasculitisfoundation.org/](http://www.vasculitisfoundation.org/)
- **Vasculitis Foundation Canada** - [www.vasculitis.ca/](http://www.vasculitis.ca/)
- **Mayo Clinic USA** - (Arizona. Florida. Minnesota) - [www.mayoclinic.org/](http://www.mayoclinic.org/)
- **Lauren Currie Twilight Foundation** - [www.thelaurencurrietwilightfoundation.org/](http://www.thelaurencurrietwilightfoundation.org/)



### Disease specific websites

- **Behçet's Syndrome Society** - phone: (helpline): 0845 130 7329 or 0845 130 7328. [www.behcets.org.uk](http://www.behcets.org.uk)
- **Churg-Strauss Association** - [www.cssassociation.org/](http://www.cssassociation.org/)
- **Henoch-Schönlein Purpura Support Group** - phone: 01733 204368 (10am-2pm) e-mail: HSP Helpdesk - [hsphelp@inbox.com](mailto:hsphelp@inbox.com)
- **Kawasaki Disease Support Group** - phone: 02476 612178 Best time to telephone: 9am - 10pm, Monday-Sunday. [www.kssg.org.uk](http://www.kssg.org.uk)
- **Microscopic Polyangiitis message boards** - [www.facebook.com/pages/Microscopic-Polyangiitis-MPA-Support](https://www.facebook.com/pages/Microscopic-Polyangiitis-MPA-Support)
- **PMRGCAuk - Polymyalgia Rheumatica and Giant Cell Arteritis** - phone 0300 111 5090 or 0300 999 5090. [www.pmrcauk.com](http://www.pmrcauk.com). email: PMRGCAuk Enquiries - [info@pmrcauk.com](mailto:info@pmrcauk.com)
- **PMR-GCA Scotland** - phone: 0300 777 5090. [www.pmrangca.org.uk](http://www.pmrangca.org.uk)
- Relapsing Polychondritis Support Group - 21 Staneway, Leam Lane, Gateshead, Tyne & Wear, NE10 8LR

### Vasculitis UK Web Discussion Groups

- **Vasculitis-UK HealthUnlocked** - A discussion group for people with vasculitis and for people with an interest in vasculitis. You can set your own levels of privacy for who sees your posts. [www.vasculitis-uk.healthunlocked.com](http://www.vasculitis-uk.healthunlocked.com)
- **Facebook (Vasculitis UK):** - Become a member of Facebook - Vasculitis UK - [www.facebook.com/groups/16351007695](https://www.facebook.com/groups/16351007695) This is a closed group where only members can see your post.
- **Facebook (Vasculitis UK Young Vasculitis):** - A closed group for young vasculitis patients between the ages of 16 and 30ish to chat to others in a safe environment. - [www.facebook.com/groups/505201572880821/?ref=ts&fref=ts](https://www.facebook.com/groups/505201572880821/?ref=ts&fref=ts)
- **Facebook (Vasculitis UK Bereavement Support):** - A closed and safe group for bereaved family and friends to chat to others who have been bereaved because of vasculitis - [www.facebook.com/groups/184007648435810/?ref=ts&fref=ts](https://www.facebook.com/groups/184007648435810/?ref=ts&fref=ts)
- **Facebook (Vasculitis UK Caring for Carers):** - A closed and safe group for those who are caring for a vasculitis patient. Chat and find help and support from others who care for a patient. [www.facebook.com/groups/162917153730920/](https://www.facebook.com/groups/162917153730920/)
- **Facebook (Vasculitis UK Music for relaxation and meditation):** - For music to help with relaxation, meditation or just to sit and close your eyes and rest for a while join. <https://www.facebook.com/groups/232118723655457/>
- **Facebook (Vasculitis UK Timeline):** - An open group with up-to-date information, news, photographs and much more. [www.facebook.com/VasculitisUK](https://www.facebook.com/VasculitisUK)

### Discussion Groups - Individual Vasculitis Diseases

"Facebook" also has a number of Discussion Groups for the individual vasculitides. Go to "Facebook" and search for the disease.

## Further information for medical professionals and patients

### Vasculitis Rare Disease Group

The Vasculitis Rare Disease Group has developed from the UK and Ireland Vasculitis registry initiative UKIVAS. The group members represent Vasculitis units across the UK and participate in Vasculitis research and care.

The Vasculitis RDG website is currently not complete but is available to view at: [RareRenal.org - Vasculitis Rare Disease Group](http://RareRenal.org - Vasculitis Rare Disease Group)

### Vasculitis and the Dental Team

An article written by John Mills, Chairman - Vasculitis UK, for the dental magazine "Vital". We are grateful to "Vital" for giving permission to reproduce the article. It can be downloaded at: [www.vasculitis.org.uk/professionals/general-information](http://www.vasculitis.org.uk/professionals/general-information)

### Approach to a patient with vasculitis

A PowerPoint presentation - "Approach to a patient with vasculitis" [www.slideworld.com](http://www.slideworld.com)

### Clinical Trials

This is an American website funded by the National Institutes of Health. All new clinical trials (for all diseases) should be registered with this organisation before starting and some of the results of the trials should be made available after completion. Some other non-trial research is also registered with the site. The information on the site is freely available and can be used to look for new clinical trials for a specific disease and where the clinical trials are taking place. Even though the site is American most European clinical trials will also be registered here. [www.clinicaltrials.gov](http://www.clinicaltrials.gov)

### European League Against Rheumatism (The) [EULAR]

The main professional organisation for European Rheumatologists' official site. The site contains links to many of their published guidelines on the management of systemic vasculitis (and many other inflammatory diseases) and on managing the side effects of drugs such as steroids and other immunosuppressants. [www.eular.org](http://www.eular.org)

### European Vasculitis Study Group (The) [EUVAS]

A collaboration of many medical professionals and researchers passionately interested in the treatment of systemic vasculitis. EUVAS has organised several major international clinical trials over the last two decades. The website is very useful containing published articles on the diagnosis, classification and management of vasculitis. The results of their clinical trials are posted to the website as are details of clinical trials underway. The website also contains links to treatment guidelines for managing systemic vasculitis. [www.vasculitis.org](http://www.vasculitis.org)

### UKVAS: a registry for patients with vasculitis in the UK and Ireland

A new project, the aim of which is to collect anonymised information about patients throughout the UK with different types of vasculitis, in a uniform way. Those involved in the project include the Vasculitis Rare Disease Working Group and is supported by Kidney Research UK and Vasculitis UK. [www.UKVAS.org](http://www.UKVAS.org)

### Diagnostic and Classification Criteria in Vasculitis Study (DCVAS)

DCVAS is a multinational observational study and aims to develop diagnostic criteria and update classification criteria in vasculitis. [www.dcvas.org](http://www.dcvas.org)

### Vasculitis Clinical Research Consortium (The) [VCRC]

The Vasculitis Clinical Research Consortium (VCRC) is an American based integrated group of academic medical centres, patient support organisations and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. The website contains links to pages on specific diseases and their treatment. It also contains links which allow patients to register their details on disease specific contact registries. [www.rarediseasesnetwork](http://www.rarediseasesnetwork)

### The Renal Association: About eGFR (Glomerular Filtration Rate)

Patient information on estimated Glomerular Filtration Rate [www.renal.org - About eGFR](http://www.renal.org - About eGFR)



## Additional reading

All articles are free access on-line unless otherwise stated. This is not an exhaustive list and more up-to-date articles are published every week. The score \* to \*\*\*\*\* is a subjective score to the possible relevance for the lay reader/patient.

**BSR and BHPR Guidelines for the management of adults with ANCA associated vasculitis (Rheumatology)**  
2014 - Revision of the 2006 Guidelines with a target audience including rheumatologists, general physicians and specialists who may come across vasculitis in the course of their work. Also specialist registrars in training, nurse practitioners dealing with vasculitis and primary care physicians. (\*\*\*\*)  
[rheumatology.oxfordjournals.org/content/early/2014/04/11/rheumatology.ket445.full](http://rheumatology.oxfordjournals.org/content/early/2014/04/11/rheumatology.ket445.full)

### Paediatric Guidelines

2012 - Vasculitis Paediatric Guidelines from the Oxford Handbook of Paediatric Rheumatology (with permission from OUP). Oxford specialist Handbooks in Paediatrics, Paediatric Rheumatology edited by Helen Foster and Paul A Brogan. (\*\*\*\*) Download at:  
[www.vasculitis.org.uk/professionals/paediatric-vasculitis-guidelines](http://www.vasculitis.org.uk/professionals/paediatric-vasculitis-guidelines)

### Recommendations on the use of Rituximab in ANCA-associated Vasculitis

2012 - Advisory document on the use of Rituximab. (\*\*\*\*) Download at:  
[www.vasculitis.org.uk/professionals/recommendations-rituximab-anca-vasculitis](http://www.vasculitis.org.uk/professionals/recommendations-rituximab-anca-vasculitis)

### General approaches to the diagnosis and management of vasculitis

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2857937/pdf/cei0160-0143.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2857937/pdf/cei0160-0143.pdf)  
2009 - A comprehensive guide to identification, cause, treatment and prognosis of all forms of vasculitis including photographic illustrations. (\*\*\*)  
Keywords: ANCA, Giant Cell Arteritis, Microscopic Polyangiitis, Wegener's Granulomatosis.

### Infection and Vasculitis

<http://rheumatology.oxfordjournals.org/content/48/5/475.full.pdf+html>  
2009 - A summary of the inter-relationship between vasculitis and infection and the physiopathological mechanisms involved in light of current knowledge from animal models. (\*)  
Keywords: Infection, Vasculitis, Autoimmune, Immune mechanism, animal model.

### Review of anti-infective prophylaxis in vasculitis

<http://arthritis-research.com/content/pdf/ar2826.pdf>  
A summary of research results available concerning patients with primary systematic Vasculitis and risk factors concerning the use of anti-infective prophylaxis. (\*\*)

### Large Vessel Diseases

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467\\_2009\\_Article\\_1312.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467_2009_Article_1312.pdf)  
2009 - An educational review of large vessel Vasculitis focussing chiefly on the disease in children. Includes some comparisons of the variations of the disease affecting different ethnic groups. (\*\*)  
Keywords: Hypertension, Idiopathic aorto-arteritis, Takayasu arteritis.

### Giant Cell/Temporal Arteritis

[www.ccjm.org/content/78/4/265.full.pdf+html](http://www.ccjm.org/content/78/4/265.full.pdf+html)  
2011 - Educational Review. Readers will suspect Giant Cell arteritis in older patients who present with temporal headache, visual changes and other pertinent symptoms. (\*\*\*\*)  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC3014829/pdf/nihms-239316.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3014829/pdf/nihms-239316.pdf)  
2008 - An article systematically reviewing the treatment options for patients with Neuro-ophthalmic and neurological complications of GCA as well as the evidence for possible adjuvant therapies for patients with GCA. (\*\*\*\*)  
Keywords: Arteritic ischemic optic neuropathy, Giant Cell Arteritis, Steroids, Stroke, Temporal Arteritis.

### Polymyalgia Rheumatica

[www.ccjm.org/content/78/4/265.full.pdf+html](http://www.ccjm.org/content/78/4/265.full.pdf+html)  
A brief mention of PMR within an educational review on GCA. (\*\*\*\*)  
<http://archinte.ama-assn.org/cgi/reprint/169/20/1839>  
2009 - A review analysing the results of 30 studies between 1957-2008. Taken from English language articles on Cochrane databases and MEDLINE the review includes a set of comprehensive tables. (\*\*)  
[www.ncbi.nlm.nih.gov/pmc/articles/PMC2762363/pdf/cia-4-391.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2762363/pdf/cia-4-391.pdf)  
2009 - A short article comparing clinical, radiological and biochemical characteristics in three cases studied of patients with diseases mimicking PMR. (\*\*)  
Keywords: Gallium-67 scintigraphy, Polymyalgia rheumatica, Pseudogout, Post-infectious polyarthritis, RS3PE syndrome.

### Takayasu's Disease

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467\\_2009\\_Article\\_1312.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855435/pdf/467_2009_Article_1312.pdf)  
2009 - An educational review of large vessel Vasculitis focussing chiefly on the disease in children. Includes some comparisons of the variations of the disease affecting different ethnic groups. (\*\*)  
Keywords: Hypertension, Idiopathic aorto-arteritis, Takayasu arteritis.

### Medium Vessel Diseases

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467\\_2009\\_Article\\_1336.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2908435/pdf/467_2009_Article_1336.pdf)

2009 - An educational review of medium-size-vessel disease focussing on the occurrence of the three types of PAN, KD and cutaneous PAN in children. (\*\*)

Keywords: Vasculitis, Polyarteritis nodosa, Cutaneous polyarteritis nodosa, Kawasaki disease, Child.

### Kawasaki's Disease

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2845298/pdf/nihms180284.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2845298/pdf/nihms180284.pdf)

2010 - Author manuscript of an acute inflammatory illness of young children, particularly in industrialized nations and predominantly in those between 6 months and 5 years of age. (\*\*\*\*)

Keywords: Coronary artery aneurysms, Cytoplasmic inclusion bodies, IgA immune response, IgA plasma cells, Inclusion bodies, Intravenous immunoglobulin, Kawasaki disease, Synthetic antibodies, Vasculitis.

### Small Vessel Diseases

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2855433/pdf/467\\_2009\\_Article\\_1317.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2855433/pdf/467_2009_Article_1317.pdf)

2009 - An educational review. The clinical manifestations of small-vessel Vasculitis in children are described and current therapies discussed. Some of the rarer entities are not considered. (\*\*)

Keywords: ANCA-associated vasculitis, Child, Henoch-Schönlein purpura, Vasculitis.

<http://onlinelibrary.wiley.com/doi/10.1002/art.23105/pdf>

2007 - A review focussing on the challenges faced by clinicians who care for patients with medium and small vessel Vasculitis. (\*\*)

Keywords: Medium/Small Vessel Vasculitis, Epidemiology, Pathogenesis, Treatment.

### ANCA Associated Vasculitides

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2893757/pdf/tcrm-6-253.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2893757/pdf/tcrm-6-253.pdf)

2010 - A review of current trends and future prospects for the management of ANCA-associated vasculitis. (\*\*\*\*)

Keywords: ANCA, Vasculitis, Treatment

**Guidelines for the management of adults with ANCA associated vasculitis** (Rheumatology 2007;46:1-11)

[www.rheumatology.org.uk/includes/documents/cm\\_docs/2009/m/management\\_of\\_adults\\_with\\_anca\\_associated\\_vasculitis.pdf](http://www.rheumatology.org.uk/includes/documents/cm_docs/2009/m/management_of_adults_with_anca_associated_vasculitis.pdf)

2007 - Guidelines with a target audience including rheumatologists, general physicians and specialists who may come across vasculitis in the course of their work. Also specialist registrars in training, nurse practitioners dealing with vasculitis and primary care physicians. (\*\*\*\*)

Keywords: Vasculitis, Guideline, Management, Cyclophosphamide.

### Microscopic Polyangiitis

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2917831/pdf/nihms202451.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2917831/pdf/nihms202451.pdf)

2010 - An author manuscript with an historical overview and epidemiology of MPA. With pathogenesis, clinical features and treatment. (\*\*)

### Churg-Strauss Syndrome (Eosinophilic Granulomatosis with Polyangiitis)

[www.discoverymedicine.com/Christian-Pagnoux/2010/03/20/churg-strauss-syndrome-evolving-concepts/](http://www.discoverymedicine.com/Christian-Pagnoux/2010/03/20/churg-strauss-syndrome-evolving-concepts/)

2010 - A French author article considering the evolving concepts of Churg-Strauss Syndrome and outlining the need for the development of new therapeutics. (\*\*\*)

### Renal Vasculitis

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2914014/pdf/1471-2369-11-12.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2914014/pdf/1471-2369-11-12.pdf)

2010 - A systematic review of 22 articles conducted to determine the benefits and harms of any intervention for the treatment of renal vasculitis in adults. (\*)

### Lung disease in ANCA associated Vasculitis

<http://chestjournal.chestpubs.org/content/136/4/1101.full.pdf+html>

2009 - An evaluation of the pathogenic and clinical aspects of pulmonary involvement in AAV and the current therapeutic options. (\*\*)

### Wegener's granulomatosis in childhood

<http://onlinelibrary.wiley.com/doi/10.1002/art.24876/pdf>

2009 - A study comparing the criteria for WG as used by the ACR with those of EULAR/PRES in a cohort of children and describe the interval to diagnosis, presenting features and initial treatment. (\*\*\*\*)

### Henoch-Schönlein Purpura

[www.aafp.org/afp/2009/1001/p697.html](http://www.aafp.org/afp/2009/1001/p697.html)

2009 - An American article reviewing information about this disease which occurs mainly in children younger than ten year old. (\*\*\*\*)

### Behçet's disease

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2967828/pdf/CDI2011-681956.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2967828/pdf/CDI2011-681956.pdf)

2010 - A Japanese review article focussing on the role of immune reactions against oral streptococci mediated by IL-12 cytokine family in the pathogenesis of ABD. (\*\*)

[www.bmj.com/content/339/bmj.b3876](http://www.bmj.com/content/339/bmj.b3876)

2009 - An article written by a Behçet's sufferer describing his experience from the first on-set to diagnosis and treatment. Includes an article from a clinician's perspective. (\*\*\*\*\*)

#### **Buerger's disease/Thrombitis obliterans**

<http://circ.ahajournals.org/content/121/16/1858.full.pdf+html>

2010 - A clinician update of this disease which affects limbs and has exposure to tobacco as a central driver to its initiation, maintenance and progression. (\*\*\*)

#### **Central Nervous System Vasculitis**

[www.ncbi.nlm.nih.gov/pmc/articles/PMC3002614/pdf/10.1177\\_1756285609347123.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3002614/pdf/10.1177_1756285609347123.pdf)

A group of 35 reviews and observations covering all aspects of central nervous system vasculitis. (\*\*\*\*)

#### **Cogan's Syndrome**

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2808685/pdf/0392-100X.29.108.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2808685/pdf/0392-100X.29.108.pdf)

2009 - An Italian case report on a rare disorder characterized by inflammatory eye disease and vestibulo-auditory symptoms primarily affecting young white adults. (\*\*\*\*)

Keywords: Hearing loss, Interstitial Keratitis, Auto-immunity, Cogan's Syndrome.

#### **Cryoglobulinemia**

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2569912/pdf/1750-1172-3-25.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2569912/pdf/1750-1172-3-25.pdf)

2008 - A review of this rare disorder which is more prevalent in Southern Europe and linked to Hepatitis C virus infection. (\*\*)

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2805946/pdf/87.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2805946/pdf/87.pdf)

2006 - Review summarising the classification and clinical and therapeutic aspects of cryoglobulinaemic vasculitis and glomerulonephritis. (\*\*)

#### **"Other" vasculitides**

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2908464/pdf/467\\_2009\\_Article\\_1327.pdf](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2908464/pdf/467_2009_Article_1327.pdf)

2009 - An educational review of relatively rare vasculitides in the common practice of paediatric nephrologists. (\*\*\*)

Keywords: Auto-inflammatory, Syndrome, Behçet's disease, Fever, Vasculitis.

#### **Primary Angiitis of the Central Nervous System**

<http://archneur.ama-assn.org/cgi/reprint/66/6/704>

2009 - A neurological review of a very rare and difficult to diagnose form of the disease. Includes present and proposed changes to diagnostic criteria. (\*\*\*\*)

#### **Pulmonary Vasculitis**

<http://radiographics.rsna.org/content/30/1/33.long>

2010 - Radiological and clinical indicators to help in the diagnosis of the various forms of vasculitis. Includes radiology images of all the different forms. (\*\*\*\*)

#### **Rheumatoid Vasculitis**

[www.ncbi.nlm.nih.gov/pmc/articles/PMC2950222/?tool=pubmed](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2950222/?tool=pubmed)

2010 - Author manuscript considering a rare but serious complication of rheumatoid arthritis which recent reports have noted is declining in prevalence. (\*\*\*\*)

Keywords: Rheumatoid Arthritis, Rheumatoid Vasculitis, Epidemiology, Biologic Therapy.

#### **ANCA Associated Vasculitides: Advances in pathophysiology and treatment**

Abstract only available on line at:

[www.ncbi.nlm.nih.gov/pubmed/20167957](http://www.ncbi.nlm.nih.gov/pubmed/20167957)

A review providing an overview of recent advances in the diagnosis and treatment of the ANCA-associated vasculitides. With useful links to many other related sites. (\*\*\*\*)

Full paper (PDF) available from John Mills (Chair) Vasculitis UK

#### **Fatigue: a primary contributor to impaired life in ANCA-associated vasculitis**

[www.rheumatology.oxfordjournals.org/content/49/7/1383.full](http://www.rheumatology.oxfordjournals.org/content/49/7/1383.full)

2009 - A population-based case-control study to examine the quality of life in ANCA-associated Vasculitis. (\*\*\*\*)

Keywords: ANCA-associated vasculitis, Wegener's granulomatosis, Microscopic polyangiitis, Churg-Strauss syndrome, Vasculitis, Quality of life, Fatigue.

#### **Alemtuzumab (CAMPATH-1H) as Remission Induction Therapy in Behçet's Disease**

[www.blackwellpublishing.com/acrmeeeting/abstract.asp?MeetingID=774&id=89925](http://www.blackwellpublishing.com/acrmeeeting/abstract.asp?MeetingID=774&id=89925)

2010 - A short retrospective review of 20 patients treated with Alemtuzumab since 1998 in Addenbrookes Hospital, Cambridge. (\*\*\*\*)

#### **Diagnosis and management of ANCA associated vasculitis**

BMJ, 2012 - bmj.com Abstract: [www.bmj.com/content/344/bmj.e26](http://www.bmj.com/content/344/bmj.e26)

2012 - A clinical review. To access the article requires you to sign up for a seven day free trial or to access the full text requires a subscription or payment.

## Research Links

**14th ANCA and Vasculitis Workshop 2009.** Lund, Sweden and Copenhagen Denmark.

[www.vasculitis.org.uk/content/downloads/14th-anca-conf-2009.pdf](http://www.vasculitis.org.uk/content/downloads/14th-anca-conf-2009.pdf)

Abstracts from the Workshop can be viewed at:

<http://onlinelibrary.wiley.com/doi/10.1111/apm.2009.117.issue-s127/issuetoc>

**15<sup>th</sup> Vasculitis and ANCA Workshop 2011** - *To be published*

**ACR: Azathioprine Bests CellCept for Vasculitis (2010)**

**Abstract from "Medpage Today"** - (Research undertaken by Cambridge University, UK)

[www.medpagetoday.com/MeetingCoverage/ACR/23227](http://www.medpagetoday.com/MeetingCoverage/ACR/23227)

**ANCA Associated Vasculitis: Are genes important** Report by Professor Lorraine Harper, Birmingham University Hospital. Vasculitis UK Newsletter/Journal, Spring 2011, page 8. Available to view at [www.vasculitis.org.uk/content/downloads/spring-2011-Newsletter.pdf](http://www.vasculitis.org.uk/content/downloads/spring-2011-Newsletter.pdf)

**Clinical trial of Rituximab as maintenance treatment of vasculitis (proposed in late 2010)**

The trial is to be the responsibility of Dr David Jayne of Addenbrookes, Cambridge. The aim is to study Rituximab as a maintenance treatment for vasculitis. For further details please contact Dr Jayne by e-mail: [dj106@cam.ac.uk](mailto:dj106@cam.ac.uk)

**Immune system and autoimmunity**

Dr Julie Williams, Birmingham University Hospital. Reported in Vasculitis UK Newsletter/ Journal. Autumn 2011 Available to view at [www.vasculitis.org.uk/content/downloads/autumn-2011-Newsletter.pdf](http://www.vasculitis.org.uk/content/downloads/autumn-2011-Newsletter.pdf)

**"Increased risk of autoimmune diseases in families with Wegener's Granulomatosis" (Abstract) - The Journal of Rheumatology—Oct 2010**

[www.jrheum.org/content/early/2010/09/27/jrheum.091280.abstract](http://www.jrheum.org/content/early/2010/09/27/jrheum.091280.abstract)

**Monoclonal antibody no better than standard treatment but Rituximab may be better for relapsing vasculitis.** Review written by Maya Anaokar on review of two articles published in the New England Journal of Medicine, 2010. Reviewed in Vasculitis UK Newsletter/Journal. Autumn 2010, page 10. Available to view at [www.vasculitis.org.uk/content/downloads/autumn-2010-Newsletter.pdf](http://www.vasculitis.org.uk/content/downloads/autumn-2010-Newsletter.pdf)

**Modern management of primary systemic vasculitis**

Ed: Richard Watts.

Clinical Medicine. Vol 7, No. 1 Jan/Feb 2007

**Ongoing laboratory research** - Dr Neil Holden, Birmingham University Hospital: **The immune system and basis of ANCA associated vasculitis and All ANCA are equal but some ANCA are more equal than others.** Reported in Vasculitis UK Newsletter/Journal, Autumn 2010, page 8/9. Available to view at [www.vasculitis.org.uk/content/downloads/autumn-2010-Newsletter.pdf](http://www.vasculitis.org.uk/content/downloads/autumn-2010-Newsletter.pdf)

**Randomized Trial of Rituximab Versus Cyclophosphamide for ANCA-Associated Renal Vasculitis** Rachel B Jones et al N Engl J Med 2010; 363:211-220 [July 15, 2010](http://www.nejm.org/doi/full/10.1056/NEJMoa0909905)

**Update from Birmingham on Cutting Edge Research into Vasculitis**

By Julie Williams [www.vasculitis.org.uk/content/downloads/cutting-edge-research-2009-birmingham.pdf](http://www.vasculitis.org.uk/content/downloads/cutting-edge-research-2009-birmingham.pdf)

**Rituximab versus Cyclophosphamide for ANCA-Associated Vasculitis,**

John H Stone et al

N Engl J Med 2010; 363:221-232 [July 15, 2010](http://www.nejm.org/doi/full/10.1056/NEJMoa0909905)

[www.nejm.org/doi/full/10.1056/NEJMoa0909905](http://www.nejm.org/doi/full/10.1056/NEJMoa0909905) (A review of this paper can be found in the Vasculitis UK Newsletter/Journal, Spring 2011 Available to view at

[www.vasculitis.org.uk/content/downloads/autumn-2011-Newsletter.pdf](http://www.vasculitis.org.uk/content/downloads/autumn-2011-Newsletter.pdf)

**Vasculitis DNA Bank for Glomerulonephritides** (as at Dec 2010)

The Vasculitis DNA bank at Birmingham, housed in the Wellcome Trust Clinical Research Facility, is still actively collecting samples from patients, spouses and relatives.

Taking part in the research will be helping the research team to establish whether genetics or possibly environmental factors are triggers in the disease process. [wtrcf@uhb.nhs.uk](mailto:wtrcf@uhb.nhs.uk), or write to: Wellcome Trust Clinical Research Facility, UHB NHS Foundation Trust, Birmingham, B15 2TH

## Book Reviews

### **'Friendly Fire' by Professor Isenberg & Dr Morrow**

A book written for the general public about the immune system. Available from the Public Library.

**Immunological & Inflammatory Disorders of the CNS: Neil Scolding.** ISBN 070623578

### **'Patient: The True Story of a Rare Illness' by Ben Watt**

The autobiography of a young musician who developed Churg-Strauss Syndrome. Published by Grove Press: New York, 1996 ISBN 0-802 13583-8

### **"Vasculitis" authors S Louis Bridges Jr and Gene V Ball**

ISBN 10: 0198570198, ISBN 13: 978-0198570196

### **'Vasculitis' Balliere's Clinical Rheumatology, vol. 11 No. 2, 1997 Editors: H. Yazici & G. Husby**

A medical textbook which aims to provide an update on vasculitis - only for those who want all the details or perhaps to provide more detailed information for your GP. Published by Balliere Tindall: London. ISBN 0-7020-2266-7 (single copy)

### **"Vasculitis in Clinical Practice" editors - Richard A Watts & David GI Scott**

ISBN: 978-1-84996-246-9 e-ISBN@ 978-1-84996-247-6 DOI: 10.1007/978-1-84996-247-6

Published by Springer Dordrecht Heidelberg London New York (Available from Amazon in paperback £34.19)

### **'Vasculitis' by the Arthritis Research Campaign (ARC)**

This booklet aims to explain what the term 'vasculitis' means, what the different types are and how it is recognised and treated. For further information regarding how to obtain a copy contact: ARC, Copeman House, St. Mary's Court, St. Mary's Gate, Chesterfield, Derbyshire, S41 7TD. Tel: 0 1246 558033 [www.arc.org.uk](http://www.arc.org.uk)

### **Wegener's Granulomatosis Booklet for the newly diagnosed:**

[www.stjames.ie/Departments/DepartmentsA-Z/I/Immunology/DepartmentinDepth/Wegeners.pdf](http://www.stjames.ie/Departments/DepartmentsA-Z/I/Immunology/DepartmentinDepth/Wegeners.pdf)

## Articles published in the Vasculitis UK Newsletter/Journals

*These articles can be viewed on the Vasculitis UK website: [www.vasculitis.org.uk](http://www.vasculitis.org.uk) To obtain a printed copy of the articles please contact John Mills or Pat Fearnside (see contact details page)*

### Spring 2014 - Issue 47

- Rituximab approved by NICE
- Physical activity in patients with GCA - Dr Joanna Robson et al, Oxford
- Potted history of the vasculitides
- Masterclass - Large vessel vasculitis
- Helping ourselves through diet - Jacky Davis

### Autumn 2013 - Issue 46

- Vasculitis in children: Dr Paul Brogan
- The UK Henoch Schonlein Purpura Nephritis Study: Dr Louise Watson
- Shingles vaccine for the over 70's
- Clinical Commissioning Groups: Dr Peter Lanyon
- Can Tai Chi be helpful?: Alice Muir
- Vasculitis: The patient experience: Dr Nina Brown
- Dedicated doctors share knowledge
- Role of Metacognitions in adapting to living with vasculitis: Helen Mayor
- MYPAN research

### Spring 2013 - Issue 45

- All ANCA aren't equal in vasculitis
- Infections in people with vasculitis
- Vasculitis and the ear
- Prednisolone and insomnia
- Vasculitis Masterclass
- Urticarial vasculitis
- Renal PatientView
- A day at Imperial College, London

### Autumn 2012 - Issue 44

- Vasculitis and the Eye
- The inflammatory actions of ANCA-activated neutrophils
- Patient reported outcome (PRO) questionnaire development for patients with ANCA-associated vasculitis
- Cogan's Syndrome
- Shingles

### Spring 2012 - Issue 43

- Diagnostic techniques in vasculitis (Report on Dr David Carruthers' presentation)
- Why do patients with ANCA Associated vasculitis experience fatigue (Report on Dr Andrew McClean's presentation)
- Fatigue a principal contributor to impaired quality of life in ANCA-associated vasculitis (AAV): Review by Maya Anaokar
- Clinical review on the diagnosis and management of ANCA-associated vasculitis (AAV): Review by Maya Anaokar
- Fertility and Vasculitis: Dr David Jayne
- Lupus, diet and lifestyle: by Ani Richardson

### Autumn 2011—Issue 42

- The immune system/autoimmunity: Dr Julie Williams
- Mycophenolate Mofetil –v– Azathioprine: Review by Maya Anaokar
- Walking the tightrope: Dr Richard Watts
- Report from AGM (Who gets vasculitis and why?): report prepared by Maya Anaokar
- About Stuart Strange, the Trust and Martin Lockwood
- Influenza vaccination

- Chickenpox and immunosuppressed people: Viv Dunstan
- Healthy eating (part 2) — High and low potassium foods: Catherine Cotter
- The enlightened patient

***Spring 2011-Issue 41***

- Healthy Eating (part 1) High and low potassium diets: Catherine Cotter
- Vasculitis and oral health: John Mills
- Blood test monitoring
- ANCA-Associated vasculitis—Are Genes Important?: Professor Lorraine Harper
- The Wellcome Trust Clinical Research Facility, Birmingham: Dr Julie Williams
- An update on Rituximab: Dr David Jayne
- Denosumab for prevention of fractures

***Autumn 2010—Issue 40***

- The Immune system and basis of ANCA associated vasculitis: Dr Neil Holden
- Can B cells and neutrophils “talk” to each other?: Dr Neil Holden
- All ANCA are equal but some ANCA are more equal than others: Dr Neil Holden
- Monoclonal antibody no better than standard treatment but Rituximab may be better for relapsing vasculitis:  
Review by Maya Anaokar

## Vasculitis videos, podcasts, webinars and slides

**Autoimmune Disease Association - Webinar** "Vasculitis in Autoimmunity: A Common Thread"  
[www.youtube.com/watch?v=HOZs3BmhNB0&mid=5260](http://www.youtube.com/watch?v=HOZs3BmhNB0&mid=5260)

### Behçet's Disease explained



To view a video explaining about Behçet's and patients speaking about their own journey with Behçet's, log onto this site, then scroll down to find the video.

[www.bhecets.com/site/pp.asp?c=bhJIJSOCJrH&b=260521](http://www.bhecets.com/site/pp.asp?c=bhJIJSOCJrH&b=260521)

/

### Immunisations and the Immunocompromised Patient

UNC Kidney Centre Director Dr Ron Falk is joined by Dr. David Weber, Professor of Medicine and Paediatrics, School of Medicine and Professor of Epidemiology, Gillings School of Global Public Health at UNC. In this recording Drs Falk and Weber discuss immunisations and the recommendations for patients whose immune systems are compromised. Dr Falk poses questions for Dr Weber that are frequently asked by patients, and discussions include general recommendations as well as specific recommendations for the flu shot, DPT, pneumonia, Hepatitis B, Hepatitis A and HPV vaccine.

[www.unckidneycenter.org/kidneyhealthlibrary/podcast\\_immune.html?mid=5123](http://www.unckidneycenter.org/kidneyhealthlibrary/podcast_immune.html?mid=5123)

**Vasculitis Medication Review.** Dr Kenneth Warrington, Associate Professor of Medicine, Mayo Clinic, USA. A video giving clips of Dr Warrington's talk on the toxicity of vasculitis medication.

[www.youtube.com/watch?v=8-tPew9Fqb8](http://www.youtube.com/watch?v=8-tPew9Fqb8)

### Vitamin D Deficiencies May Impact Onset of Autoimmune Lung Disease

**Science Today** (Jan. 5, 2011) — A new study showing that vitamin D deficiency could be linked to the development and severity of certain autoimmune lung diseases.

[www.sciencedaily.com/Releases/2011/01/110104064020.htm](http://www.sciencedaily.com/Releases/2011/01/110104064020.htm)

**Slides presenting complete review on ANCA vasculitis and Anti GBM disease** - Dr Peter Bryan Schrier

[www.slideshare.net/kenar78/anca-vasculitis-anti-gbm?from=share\\_email](http://www.slideshare.net/kenar78/anca-vasculitis-anti-gbm?from=share_email)

**Video (slides presentation + audio) by Dr Leonard Calabrese reviewing novel therapies to treat vasculitis, specifically rituximab for ANCA-associated vasculitis**

[www.youtube.com/watch?v=ELP05fWned0](http://www.youtube.com/watch?v=ELP05fWned0)

**Video (slides presentation + audio) by Dr Reginald Obi, Rituximab versus Cyclophosphamide for ANCA+ vasculitis: A review of RAVE and RITUXIVAS Trials**

[www.youtube.com/watch?v=h9D2kRMY8YE](http://www.youtube.com/watch?v=h9D2kRMY8YE)





## Vasculitis Support Groups

<b>Beds, Bucks &amp; Herts</b>	<b>Janine Davies Christine Lee</b>	01525 372733 01480 869162	<a href="mailto:family.davies@btinternet.com">family.davies@btinternet.com</a>
<b>Cambridge</b>	<b>Lesley Noblett</b>	07765 897780	<a href="mailto:lnoblett@gmail.com">lnoblett@gmail.com</a>
<b>Canterbury area *</b>	<b>Margaret McGrath *</b>	01227638469	<a href="mailto:margaretmcgrathfmsj@yahoo.com">margaretmcgrathfmsj@yahoo.com</a>
<b>East Kent *</b>	<b>Brian Hart *</b>	01227 369774	
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