



Vasculitis

A Guide for Patients



What is Vasculitis?

Vasculitis is a rare disease which affects about one in 40-50,000 people each year. Vasculitis means inflammation causing damage to the walls of blood vessels. This damage can cause the vessel to partially or completely block off, cutting off the blood supply to organs and preventing them from working normally. Vasculitis can affect large, medium or small blood vessels so the symptoms of vasculitis can appear anywhere in the body.

Your immune system normally protects you from harmful bacteria and viruses. Vasculitis is an *autoimmune* illness. An autoimmune response is the immune system mistaking "self" (you) as "non-self" (stranger). The body's own immune system starts to attack the cells lining the blood vessel wall injuring them.

What causes vasculitis?

Doctors and scientists are coming closer to understanding what causes vasculitis but a lot remains to be discovered. It does not seem to be hereditary. Possible trigger factors include viral infections, environmental exposures or a combination of these and other factors, but no one knows for sure.

There are several types of vasculitis and they are all related. You may have heard your Doctor talk about you having an "ANCA" autoantibody in your blood.

What is an Antibody? Antibodies are proteins normally generated by the immune system to protect us against invading viruses and bacteria.

What is an Autoantibody? An autoantibody is an abnormal antibody that mistakenly targets "self" (you) instead of a bacteria or a virus.

What are ANCA? ANCA are a type of autoantibody found in many patients with small-vessel vasculitis. ANCA are used to help in the diagnosis of small-vessel vasculitis.

ANCA stands for Anti-Neutrophil Cytoplasmic Antibody. A Neutrophil is a type of white blood cell that normally helps fight infection.

You may have one of two types of ANCA:

1. An ANCA directed against a part of neutrophils known as myeloperoxidase. It is sometimes referred to as “MPO-ANCA” or “P-ANCA”
2. An ANCA directed against a part of neutrophils known as proteinase 3. It is sometimes referred to as “PR3-ANCA” or “C-ANCA”.

Vasculitis can occur even in the absence of an ANCA and while measuring the ANCA level is useful for monitoring the disease it is not 100% accurate at telling us whether your disease is active or not.

Some patients may have vasculitis without having a detectable ANCA in their blood and you can also have an ANCA without vasculitis.

The types of vasculitis described in this booklet are:

- Microscopic Polyangiitis
- Granulomatosis with Polyangiitis (Wegener’s Granulomatosis)
- Eosinophilic Vasculitis (Churg-Strauss Syndrome)

Who gets vasculitis?

Vasculitis occurs for the first time in 10-20 people every year in Cork and Kerry. It affects mostly middle age to older people with the average age being in the early 60s. However, young children and very elderly people can develop the disease also.

Vasculitis affects men and women equally.

What types of vasculitis are there?

Many different parts of the body may be affected. Parts of the body that might be affected include the following:

- Kidney
- Joints
- Lung
- Skin
- Sinuses / Nose
- Nerves
- Gastrointestinal tract (Gut)
- Eyes

How long will the vasculitis last?

Your vasculitis may be a short-term illness (months) and cured by treatment. However, it may develop into a long term (“chronic”) disease that you will have to manage for the rest of your life. If your disease is chronic, you may have long periods when it seems to go away. This is called a “remission”. When it recurs this is called a “relapse”. Because there is always the possibility of it returning, you will need to be checked by your doctor on a regular basis.

What are the symptoms of vasculitis?

The symptoms of vasculitis are easily confused with other illnesses, so the disease often goes unrecognised for a long time. Many people first have flu-like symptoms with fever, body aches, pain in joints and muscles, reduced appetite and weight loss. More people become sick in the late Autumn, winter or early spring than in other seasons. You can feel very tired, and may need to sleep for many hours at a time even after your

treatment has begun. Your joints may ache and even swell. You may have all or some of the symptoms or you may have different ones at different times.

It is important to be attentive for new or worsening symptoms that might develop, even while you are on treatment. These may suggest a disease relapse, be a side effect of treatment or be co-incidental. Discuss them with your Doctor.

The following is a list of symptoms for each area of the body:

- Ear: hearing loss, pain, a feeling of fullness
- Eye: red eyes that hurt, blurry vision, headaches
- Gastrointestinal tract: pain in your stomach or blood in your bowel motion
- Joints: pain and/or swelling to different parts of your body. Only a few joints may be involved or many.
- Kidney: urine turns brown, tea-colored or red – meaning a leakage of blood through the inflamed kidney into the urine
- Lung: a cough - this can be mistaken for pneumonia. If you cough up blood you must see your doctor right away.
- Nose: a “runny nose” that becomes worse, usually caused by sinus drainage. You may also have pain in your nose, recurrent nose bleeds or have trouble breathing through your nose.
- Trachea: shortness of breath
- Skin: red or purple spots that show up on your skin, ulcers, itching, hives or a rash.

Is vasculitis contagious?

No. Vasculitis cannot be caught by friends or family.

Is vasculitis hereditary?

Vasculitis does not seem to run in families.

What types of tests and examinations are done to diagnose vasculitis?

Your doctor will use a combination of your medical history, a physical examination, blood tests, X-rays and possibly a biopsy to diagnose vasculitis. Your doctor will focus on the body systems that are most commonly affected and examine those specific areas.

A biopsy of the affected area may be needed to help your doctor to make a diagnosis and decide what type of treatment will be best for you. A biopsy means that a small piece of tissue is taken so it can be looked at under a microscope. A specialist doctor known as a Pathologist will examine the biopsy for evidence of damage to blood vessels to help make the diagnosis of vasculitis.

Sometimes your doctor may refer you to another doctor who specialises in the treatment of specific organs.

- A blood test will be used to find out if you have an ANCA.
- Blood and urine tests will be used to tell if the vasculitis is in your kidney.
- Your doctor may listen to your lungs and get a chest x-ray or a CT scan to check for problems in your lungs.
- Your doctor may send you to an Ear, Nose & Throat (ENT) specialist to look at your entire upper respiratory tract using a special scope.
- Your doctor may send you to an Eye Doctor (Ophthalmologist) specialist to look examine your eyes.

Do I need to see a specialist before I begin treatment?

Yes, because vasculitis is so rare, it is important to see one or more experts in this field who can diagnose your disease, recommend treatment and follow up with you on a long-term basis.

Vasculitis can affect many organ systems in your body. So you may need to be cared for by several specialists who can coordinate your care.

What type of treatment will I be receiving?

Your doctor will be using the results from blood tests and other tests to decide which treatment will be best for you. The goal of treatment is to quickly stop the inflammation that can cause organ damage while not over-treating and causing infections or other side effects.

Treatment is a balancing act: too little and symptoms of the disease may not come under control or flare later, too much and infections and other complications can occur.

The treatment you receive is based on:

- The type of vasculitis you have
- The severity of your disease
- How many of your organ systems are affected

What do I need to do to prepare for treatment?

Being told you have a rare and serious disease can be stressful and overwhelming. Your Doctor or Nurse Specialist will be giving you lots of information about the treatment options. While you are learning about these choices and what decisions to make, it is important to bring a family member or friend to your appointments. That allows someone else to help you take notes and ask questions so that you don't have to remember everything you're being told.

Here are some questions you may want to ask your doctor and other health team members.

- What is my diagnosis?
- What stage is my disease?
- What are my treatment do you recommend for me? Why?
- What are the chances that the treatment will be successful?
- What are the risks and side effects of treatment?
- How long will my treatments last?
- Will I have to change my normal activities?

There are note pages at the end of this booklet to write down the answers to your questions as you talk to your Doctor.

What are the types of treatment used for my disease?

There are several treatment options for your disease. These are often used in combination in an attempt to control the disease while minimising any harmful or unpleasant side effects.

- Corticosteroids (Prednisolone / Methylprednisone). Commonly known as "steroids" these may be given intravenously or in pill form. Corticosteroids work by controlling inflammation and suppressing the immune system.
- Immunosuppressive drugs such as cyclophosphamide, methotrexate, azathioprine (Imuran), mycophenolate mofetil (CellCept / Mycolat) or Rituximab may be given. They suppress the immune system and help kill the immune system cells that cause damage to the blood vessels.

- Antibiotics such as trimethoprim/sulfamethoxazole (Septrin) may help prevent relapses, especially in the nose.
- Plasmapheresis, also known as plasma exchange is a method of directly removing ANCA antibodies from blood. This is performed in the hospital and may help in more severe cases.

What are the side effects of these medicines?

Side effects can be a part of any treatment program and will affect each person differently. Other, less common side effects not listed here may also occur. Some of the more common side effects for each drug are:

Corticosteroids

- Sleeplessness – depending on the dose, you may find it hard to sleep. Your doctor can prescribe medicines to help you sleep. This side effect usually goes away as the steroid dose decreases
- Weight gain – you may gain weight when you are on larger doses of steroids, especially in your face. To help avoid weight gain, eat lots of fruits and vegetables and keep a supply of ready-to-eat, healthy snacks available to help satisfy your cravings
- Hair growth – you may notice an increase in hair growth on your face and body. This will return to normal as your steroid dose decreases.
- Mood swings – your mood may go from feeling very happy to very sad or you may feel more irritable or become easily upset. Knowing that you will be experiencing this may help you to develop your own coping strategies. Talking with a friend, family member or counselor may also help.

Cyclophosphamide

- You may have a drop in your white blood cells. This means you are at a higher risk for infection, and should try to stay away from people who are sick, for example someone with a fever or the flu. You may also have a fall in your red blood count (anaemia) and feel more tired than usual.
- Nausea and vomiting – anti-nausea medicines can relieve your symptoms

- To help prevent damage to your bladder, try to drink lots of fluids on the day of your infusion.
- Your hair may thin as a result of taking the medicine, but will grow back after you finish.

Azathioprine

- You may have a drop in your white blood cells. This means you are at a higher risk for infection, and should try to stay away from people who are sick, for example someone with a fever or the flu. You may also have a fall in your red blood count (anaemia) and feel more tired than usual.
- You may bleed more easily so you will need to be very careful with razors, toothbrushes, knives, and nail cutters
- Sometimes this medicine can cause nausea or vomiting. Taking it after meals and at bedtime may help. Anti-nausea medications can also relieve your symptoms.

Mycophenolate Mofetil

- You may have a drop in your white blood cells. This means you are at a higher risk for infection, and should try to stay away from people who are sick, for example someone with a fever or the flu. You may also have a fall in your red blood count (anaemia) and feel more tired than usual.
- Nausea, vomiting, diarrhoea. Taking the medicine in 3 divided doses rather than twice a day may decrease the nausea or diarrhoea (always consult your doctor before changing the way you take your medicine).

Rituximab:

- This is given as an intravenous infusion with spaced doses several weeks apart.
- Some people may experience allergic side effects to the infusion. These may include low blood pressure, shortness of breath, and skin rashes.
- You may have a drop in your white blood cells. This means you are at a higher risk for infection, and should try to stay away from people who are sick, for example someone with a fever or the flu.
- Other side effects related to the infusion include abnormal heart rhythm, nausea, and vomiting.

Septin / Co-Trimoxazole:

- Increased sun sensitivity can occur, so it is important to avoid the sun when possible, and wear sunscreen and protective clothing when out in the daylight.
- Contact your doctor immediately if you develop an allergic reaction such as difficulty breathing, tightness of the chest, swelling of the eyelids, face or lips, rash or hives

Methotrexate:

- You may have a drop in your white blood cells. This means you are at a higher risk for infection, and should try to stay away from people who are sick, for example someone with a fever or the flu. You may also have a fall in your red blood count (anaemia) and feel more tired than usual.
- Nausea and vomiting – anti-nausea medicines can relieve your symptoms
- Mouth sores
- Bleeding from gums, nose
- Blood found in urine or bowel movement

It is very important to tell your doctor if you are unable to take your medication for whatever reason.

If you develop a fever while on any of these medications it is important to seek immediate medical advice.

Please bring an up to date list of your medications with you to every clinic visit.

What will happen after treatments are finished?

Following your first treatment phase, you may have a long period of remission. Most patients are in remission after 3-6 months of treatment.

You will need to visit your doctor on a regular basis to monitor you for side effects of the medicines you are taking and to make sure you are disease-free.

What if I have a relapse or a flare?

A relapse or flare is typical for many chronic illnesses. Almost one in two patients with vasculitis will suffer a relapse of the illness having first enjoyed a period of remission.

You may have tests to make sure your illness is a relapse of vasculitis and not some other illness. If it is a relapse, you and your doctor will choose the best way to treat it. Your therapy may involve taking some of the same medicines you took when you were first treated.

What is the outlook for patients with vasculitis?

The outlook for vasculitis patients has greatly improved with therapy. Patients now routinely go into remission and live long, productive lives. Doctors continue to look for the perfect balance of therapy, meaning one that is highly effective and less toxic.

What types of support systems are there for patients with vasculitis?

Having a rare, chronic disease can be difficult to handle on your own. Sometimes talking to someone about your concerns may help. A friend, family member or a counselor can listen to your thoughts and concerns and help you find ways to cope.

The resources page on the next page of this booklet has a listing of organisations that provide education and support to patients with vasculitis.

Resources

Cork University Hospital (CUH) – The CUH Department of Renal Medicine's is an active participant in the UK and Ireland Vasculitis Registry (UKIVAS). We are involved in several clinical trials in the field of vasculitis aimed at improving the treatment options available to patients with this condition. If you are interested in participating in a clinical trial condition please speak with your Doctor.

Vasculitis Awareness Ireland (VIA) –VIA is an all-Ireland support group for patients with vasculitis.
Contact: vasculitisireland10@gmail.com

Irish Kidney Association (IKA) - The Irish Kidney Association is a charitable voluntary organisation dedicated to meeting the needs of kidney patients and their families and carers.
Internet: www.ika.ie

Vasculitis UK: - A UK based charity whose goal is to support those suffering from vasculitic diseases and their families by providing information and advice.
Internet: www.vasculitis.org.uk/

The Vasculitis Foundation - (formerly the Wegener's Granulomatosis Association) a non-profit US organization dedicated to providing emotional and informational support to patients with vasculitis, to assisting them and their families in understanding the disease and to supporting research into the cause and cure of these diseases.
Internet: www.vasculitisfoundation.org

Contacts:

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- Renal Ward (4C) Cork University Hospital 021 4920889
- Renal Department Secretary (9am-5pm) 021 4234018

