UNC KIDNEY CENTER

ANCA Vasculitis

What is Vasculitis?

VASCULITIS is inflammation ("-itis") of blood vessels (vasculum = vessel in Greek). INFLAMMATION is caused by white blood cells attacking something in a tissue. When "-itis" is at the end of a diagnosis, it means that inflammation is involved in the disease. For example, vasculitis (inflammation of any type of vessel), arteritis (inflammation of arteries), glomerulonephritis (inflammation of glomerular capillaries), arthritis (inflammation of joints), dermatitis (inflammation of skin), etc. There are many different causes of vasculitis. Some vasculitis is caused by infection of vessels (as in Rocky Mountain spotted fever). Another cause of vasculitis is AUTOIMMUNITY.

What is Autoimmune Vasculitis?

AUTOIMMUNE VASCULITIS is caused when the immune system causes vasculitis. Normally, the immune system defends against foreign invaders (such a infectious microorganisms, germs) by causing white blood cells (inflammatory cells) to attach the invaders and cause inflammation. In an autoimmune disease, the immune system causes white blood cells to attack ones own self (autos = self). In different autoimmune diseases, white blood cells attack different cells and tissues of the body. In AUTOIMMUNE VASCULITIS white blood cells attack ones own blood vessel walls causing inflammation of the vessels (vasculitis).

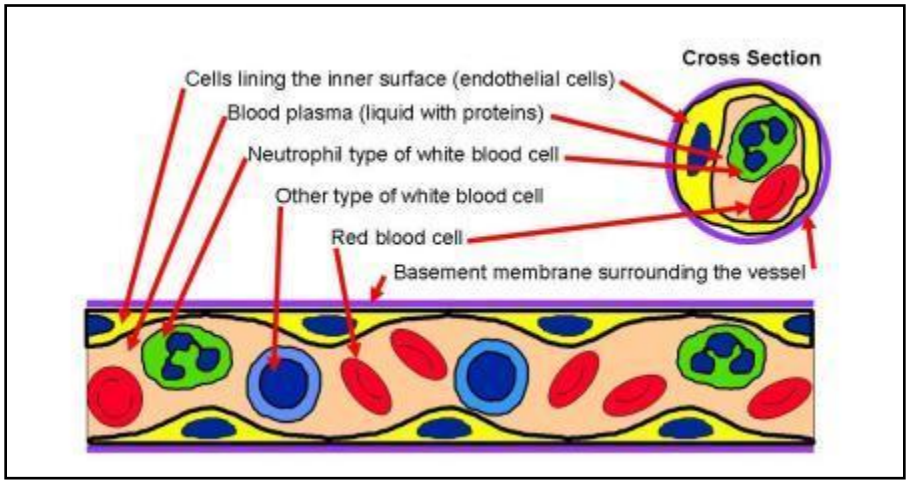
What is ANCA Vasculitis?

ANCA vasculitis is a type of autoimmune vasculitis caused by autoantibodies. Normal antibodies are molecules in the blood that are produced by the immune system to attack foreign invaders, such a infectious microorganisms (germs).

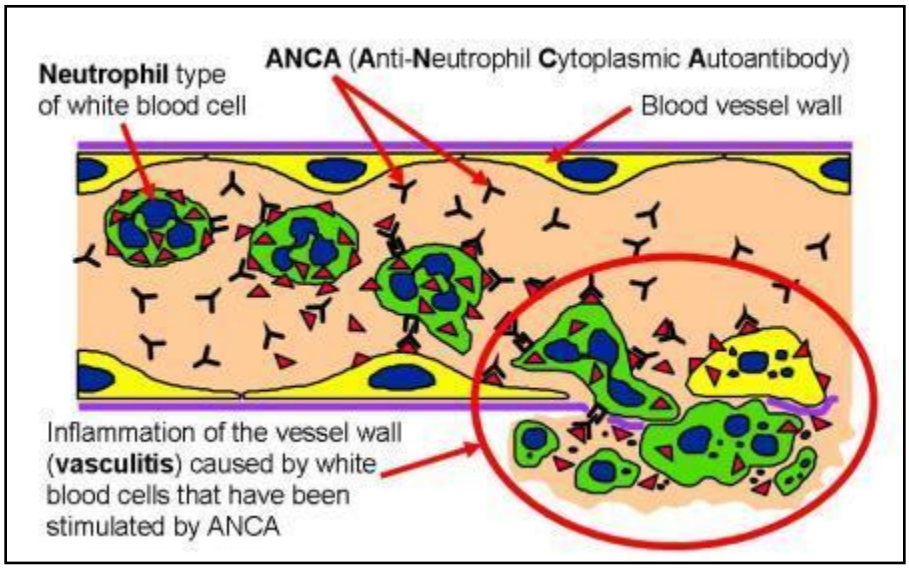
AUTOANTIBODIES are abnormal antibodies that attack ones own cells and tissues (autos = self). ANCAs are autoantibodies that attack the inside (cytoplasm) of a certain type of white blood cells called neutrophils (**ANCA** = **A**nti-**N**eutrophil **C**ytoplasmic **A**utoantibody). When ANCAs attack these white blood cells (neutrophils) they cause the white blood cells to attack the walls of small vessels in many different tissues and organs of the body resulting in vasculitis. For example, vasculitis in the skin causes red spots (purpura), in the lungs or nose causes bleeding (hemorrhage), in the nerves causes tingling or weakness (neuropathy), in the eyes causes redness and itching, and in the kidneys causes leaking of blood and protein into the urine (hematuria and proteinuria) and kidney failure.

Blood Vessel and Blood Cells

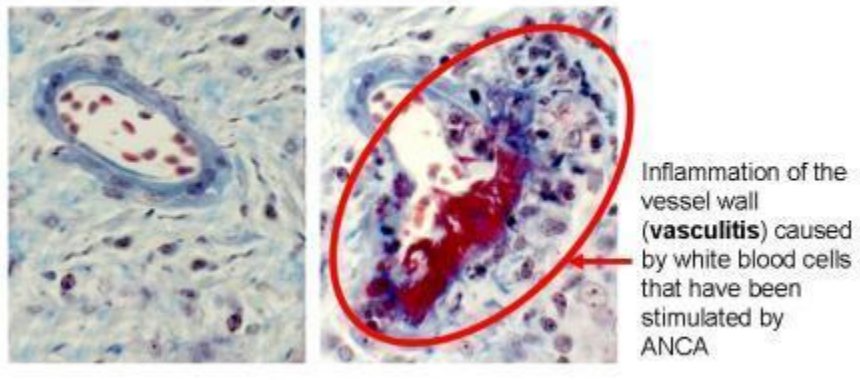
Below is a diagram of a cross section and a length-wise section of a SMALL BLOOD VESSEL. The blood vessel contains blood that is composed of red cells and white cells floating in a liquid called plasma. The inner surface of the vessel is lined by cells (endothelial cells) that sit on a thin membrane.



ANCA VASCULITIS is caused when ANCAs attack white blood cells (neutrophils). This causes the white blood cells to attack vessel walls resulting in vessel wall inflammation (vasculitis).



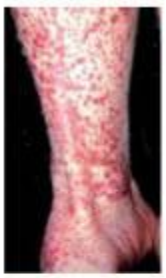
Shown below on the left is how a normal small vessel would look under the microscope in a biopsy specimen. On the right is how the vessel looked after it was injured by ANCA VASCULITIS. Compare this to the earlier diagram to understand what has happened.



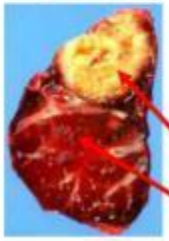
What are the different types of ANCA vasculitis?

In different people, ANCA vasculitis attacks different organs and may be accompanied by different other disease symptoms that are used to make the diagnosis of a specific form of ANCA vasculitis.

- **Renal limited vasculitis or ANCA glomerulonephritis**: the vasculitis is causing glomerulonephritis with no involvement of other organs.
- **Microscopic polyangiitis**: the vasculitis is causing injury to blood vessels in multiple tissues at the same time, such as kidneys, skin, nerves, and lungs.
- **Wegener's granulomatosis**: the vasculitis is accompanied by a nodular form of destructive inflammation called granulomatous inflammation. This often affects the lung, sinuses, nose, eyes or ears.
- **Churg-Strauss syndrome**: the vasculitis is accompanied by a granulomatous inflammation and the patient also has asthma



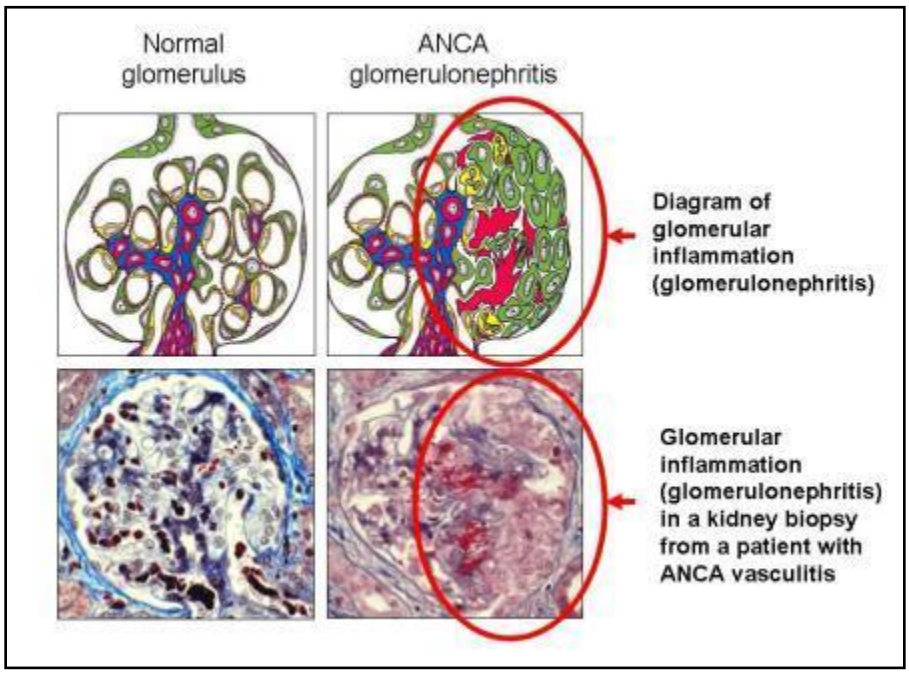
Multiple red spots on the lower leg caused by vasculitis in the skin. The vasculitis has punched holes in the vessel walls causing local bleeding into the skin (this is called **purpura**). Purpura results from many types of vasculitis, including microscopic polyangiitis, Wegener's granulomatosis and Churg-Strauss syndrome.



The piece of lung at the left came from a patient with Wegener's granulomatosis. It has been sliced open so you can see the inside. The yellow nodule (**granuloma**) was about 3 inches in diameter.

What is ANCA Glomerulonephritis?

ANCAs can cause white blood cells (neutrophils) to attack the small blood vessel (capillaries) in the filtering units (glomeruli) of the kidney. This inflammation of glomeruli is called GLOMERULONEPHRITIS. This word means inflammation (-itis) of the glomeruli (glomerulo-) in the kidney (nephros). The destruction of the glomerular capillaries causes blood and proteins to spill into the urine (hematuria and proteinuria) and causes the kidney to stop functioning (kidney failure). Glomerulonephritis often occurs in patients with ANCA disease.



Who Gets Vasculitis?

Vasculitis is very rare, occurring in 1 out of 50,000 people. The average Vasculitis patient is 55 years old, though very young children and elderly people can also develop Vasculitis. Vasculitis affects whites more than minorities, and affects men and women equally.

What Does it Affect, and What are the Symptoms?

Vasculitis can affect any part of the body the chart below names parts of the body affected by vasculitis and the doctor who specializes in that body part:

Affected Body Part	Doctor
Kidney	Nephrologist

Joints	Rheumatologist
Lungs	Pulmonologist
Skin	Dermatologist
Sinus/Nose	ENT
Trachea	ENT
Nerves	Neurologist
Gastrointestinal Tract	Gastroenterologist
Eyes	Othalmologist

The symptoms of vasculitis include flu-like symptoms including fever, body aches, joint and muscle pain, reduced appetite, and weight loss. Symptoms can also appear in the particular body part that is affected by the disease. The chart below explains some common symptoms that are specific to the body part that is affected.

Affected Body Part	Symptoms
Kidney	Brown, tea-colored urine from blood leaking into the urine
Joints	Joint pain or swelling
Lungs	Heavy cough, often mistaken for pneumonia, coughing up blood
Skin	Red or purple spots , itching, hives, rash
Sinus/Nose	Runny nose that get worse and worse, nose pain, trouble breathing through nose
Trachea	Shortness of breath
Gastrointestinal Tract	Stomach pain, or blood in stool
Eyes	Red, painful eyes, blurry vision, headaches
Ears	Hearing Problems or Hearing Loss

How Long Does Vasculitis Last?

Vasculitis can be a short-term illness that is cured by treatment, or it can be a long-term disease that never completely goes away. Many people with long-term vasculitis live long, healthy lives and enjoy long periods when the disease has few or no symptoms; however, regular check-ups are very important for patients who have been diagnosed with vasculitis because there is always a risk that the disease will return.

What Types of Tests Detect Vasculitis?

Doctors use a combination of lab tests and physical exams to diagnose vasculitis. These tests usually focus on the area of the body that is affected. Many times patients need specialists to examine a specific area. Below are some tests commonly used to diagnose vasculitis:

- A blood test to check for ANCA
- Blood and urine tests to check to see if vasculitis is affecting the kidneys
- Chest x-rays or CT scan to check for lung problems
- Endoscopic exam by an ENT to check the ears, nose, and throat
- A small tissue sample, called a biopsy, may be taken from an affected area for a lab to examine more closely

How is it Treated?

The treatment of vasculitis can be complex, and should always involve specialists for each affected body system.

Doctors use a combination of approaches to try to reduce the symptoms of the disease without causing dangerous or uncomfortable side effects from the treatment. The goal of the treatment is to reduce the swelling to protect organs from damage without compromising the immune system and allowing other infections to spread.

Exact treatment depends on:

- The type of vasculitis
- The severity of the disease
- How many organ systems are affected

Treatment options include:

- Corticosteroids: prednisone, methyl prednisone, or Medrol) A type of steroid that can be given by IV or in pill form and help reduce swelling and slow down the autoimmune response in vasculitis.
- Immunosuppressive Drugs: (cyclophosphamide, cyclosporine, azathioprine, mycophenolate mofetil, or Rituximab) Drugs that reduce the immune system's tendency to attack itself.
- Antibiotics: (trimethoprim/sulfamethoxazole) Drugs that kill foreign infections that can flourish in the nose, or in patients with a weak immune system from immunosuppressive treatment.
- Plasmapheresis: A treatment that removes the ANCA from the blood. This treatment is usually reserved for patients with bleeding in the lungs or kidney failure.

What Should I do if I have Vasculitis?

Learning that you have a rare chronic disease can be scary, stressful, and overwhelming. Healthcare providers will be sharing lots of information about your condition and the treatment options. One of the most helpful things you can do is to have a trusted friend or family member with you at your appointments. Ask them to take notes and help you ask questions. This will reduce the stressful worry that you might forget a question, or not remember some important information your doctor tells you.Here are some questions that many patients find helpful to write down and ask their doctor:

- What is my diagnosis?
- What stage is my disease?
- What are my treatment choices?
- Which option do your recommend?
- Why?
- What are the chances that the treatment will be successful?

- What are the risks and side effects of treatment?
- How long will the treatment last?
- Will I have to change my normal activities?
- What will treatment cost?

Kidney Transplant in ANCA Vasculitis

In some patients, even with treatment, involvement of the kidney with ANCA vasculitis will lead to kidney failure. Fortunately, kidney transplant is a treatment option for these patients.

For some general information about kidney transplant, visit the UNC Kidney Center web site.

Will the ANCA disease come back in my kidney transplant?

About 10-15% of patients will have symptoms of ANCA disease after a kidney transplant, called recurrent disease. Not all episodes of recurrence affect the kidney, but some will. It is important that ANCA disease be in remission prior to getting a kidney transplant – this reduces the chance that the ANCA disease will come back after the transplant.

Generally, ANCA disease recurrence will occur between 2.5 and 4 years after transplant, but can occur any time.

Is there any treatment for ANCA disease that comes back in a transplant?

There are treatments, which are similar to those for patients with ANCA disease in their original kidneys. They include corticosteroids and immunosuppressive drugs (different from the ones taken to prevent kidney transplant rejection).

If the ANCA disease comes back, will it cause me to lose my kidney transplant?

Recurrent ANCA disease can cause loss of a transplanted kidney, but it is not common. If you look at all patients with ANCA disease 10 years after their transplant, about 8% of them will have lost their kidney due to recurrent ANCA disease.

If you have ANCA disease and get a kidney transplant, you can expect a similar life span and kidney transplant survival as the average transplant patient who has kidney failure from another disease.

The Vasculitis Foundation

The Vasculitis Foundation (VF, formerly the Wegener's Granulomatosis Association) was established to alleviate the isolation that patients and their families experience when these rare life-threatening diseases affect them. They provide support, awareness, and research for all types of vasculitis. Website: http://www.vasculitisfoundation.org

The local web site for the North Carolina/Raleigh Vasculitis Support Group can be found at http://ncvasculitissupportgroup.memberlodge.org

E-mail: vf@vasculitisfoundation.org

Clinical Trials

ClinicalTrials.gov: provides regularly updated information about federally and privately supported clinical research in human volunteers. ClinicalTrials.gov gives you information about a trial's purpose, who may participate, locations, and phone numbers for more details.

Internet: www.clinicaltrials.gov

CenterWatch Clinical Trials Listing Service: this site provides general information about clinical trials, includes listings of thousands of industry-sponsored clinical trials that are actively recruiting patients in the U.S. and internationally.

Internet: www.centerwatch.com