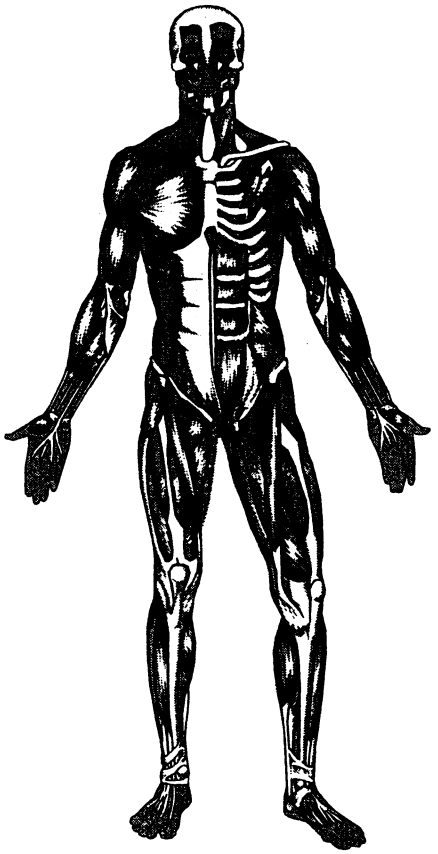
What is small vessel vasculitis?

Vasculitis is an inflammation of blood vessels in the body which causes damage to vessel linings. This damage can cause the vessel to partially or completely close, blocking off the blood supply to organs and tissues and preventing them from functioning normally. Symptoms of vasculitis can appear anywhere in the body.

Vasculitis is an autoimmune response. This means that your body's immune system turns against itself. An autoimmune response is the immune system mistaking "self" (you) as "non-self" (stranger).

The types of vasculitis described in this booklet are:

- Microscopic Polyangiitis (or Polyarteritis)
- Wegener's Granulomatosis
- Churg-Strauss Syndrome
- Organ-Limited Disease – vasculitis affecting only one organ or body site.



What causes vasculitis?

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

There are several types of small-vessel vasculitis and they are all related. Now an animal model proves that some types of ANCA can cause vasculitis, with new research in this area currently underway.

What are ANCA? ANCA cause neutrophils and monocytes (white blood cells) to damage blood vessels. ANCA are autoantibodies found in small-vessel vasculitis: Anti-Neutrophil Cytoplasmic Antibodies. These words mean that there is an antibody to the cytoplasm of neutrophils. ANCA are present in several types of small-vessel vasculitis, including microscopic polyangiitis, Wegener's Granulomatosis and Churg-Strauss syndrome.

Let's look at the meaning of these words:
Anti-means "against."

Neutrophils are a type of white blood cell containing granules filled with potent chemicals that fight infection. These chemicals play a key role in acute or inflammatory reactions.

Cytoplasmic refers to the part of the cell outside the nucleus or center of the cell.

Autoantibodies are proteins secreted by a type of immune cell that recognizes foreign substances.

ANCA are used to help in the diagnosis of small-vessel vasculitis. ANCA react to two chemicals inside normal neutrophils. These two chemicals are called myeloperoxidase (the protein that makes pus green) and proteinease 3 (an enzyme that chews up elastic tissue).

You may have one of two types of ANCA:

1. ANCA directed against myeloperoxidase- called myeloperoxidase ANCA which is sometimes referred to as "MPO-ANCA."

2. ANCA directed against proteinease 3-called proteinease 3 ANCA which is sometimes referred to as "PR3-ANCA."

Who gets vasculitis?

Vasculitis occurs in 20 people per million. It affects mostly middle age to older people with 55 being the average age. However, young children (age 2) and people age 90 or older have been known to have the disease as well. It occurs more frequently in the white population than in minority populations. 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 (the type of doctor that usually treats each symptom is listed in parentheses):

- Kidney (Nephrologist)
- Joints (Rheumatologist)
- Lung (Pulmonologist)
- Skin (Dermatologist)
- Sinus/Nose (Ear, Nose, and Throat – ENT)
- Trachea (ENT)
- Nerves (Neurologist)
- Gastrointestinal tract (Gastroenterologist)
- Eyes (Opthamologist)

How long will the vasculitis last?

Your vasculitis may be a short-term illness and cured by treatment. Or, it may be a 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 "remission". 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?

Most 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 fall, winter or early spring than in other seasons. You can feel tired, and may need to sleep for many hours at a time. 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.

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

- Ear: hearing problems or hearing loss
- Eye: red eyes that hurt, blurry vision, headaches
- Gastrointestinal tract: pain in your stomach or blood in you stool
- 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 – 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 or have trouble breathing through your nose.
- Trachea: shortness of breath
- Skin: red or purple spots that show up on your skin, itching, hives or a rash.

Is vasculitis contagious?

No. Vasculitis can not be caught by other people.

Is vasculitis hereditary?

Vasculitis does not seem to run in families.

Is vasculitis similar to AIDS?

No. Vasculitis is the opposite of AIDS because the immune system is overactive in vasculitis and the immune system is underactive in AIDS.

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

Your doctor will use a combination of laboratory tests and physical examinations to diagnose vasculitis. Your doctor will focus on the body systems that are most commonly affected and examine those specific areas. Sometimes your doctor may refer you to a doctor who specializes (a "specialist") in the treatment of specific organs.

- A blood test will be used to find out if you have ANCA.
- Blood and urine tests will be used to tell if the vasculitis is in your kidney. If so, you may be referred to see a nephrologist (kidney doctor) for more tests.
- Your doctor may listen to your lungs and get a chest x-ray or a CT scan to check for problems in your lungs. You may be referred to a pulmonologist (lung doctor) for more tests.
- Your doctor may send you to an ENT (Ear, Nose & Throat) specialist to look at your entire upper respiratory tract using a special scope.
- 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.
- Other tests may be performed and you may be referred to doctors with other specialties for these tests.

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 specialists 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 treatment.

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 stop the inflammation that can cause organ damage while not over-treating and causing infections or other problems that can occur from taking many of the prescribed drugs. Treatment is a balancing act: too little and symptoms of the disease (flares) will occur, 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 disease can be stressful and overwhelming. Your health care team 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 choices? Which 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?
- What will my treatments cost?

There are note pages at the end of this booklet to write down the answers when you talk to your doctor.

What are the types of treatment used for my disease?

You may receive several medicines for your disease.

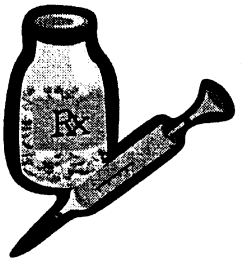
Corticosteroids (Prednisone, methylprednisone or medrol) may be given intravenously (IV) or in pill form and are similar to a hormone that is produced in the kidneys. Corticosteroids work by controlling inflammation and suppressing the immune system.

Immunosuppressive drugs such as cyclophosphamide (Cytoxan), cyclosporine, azathioprine (Imuran), mycophenolate mofetil (CellCept) or Rituximab may be given in pill form or intravenously (IV). They suppress the immune system and help kill the cells that cause inflammation in the blood vessels.

Antibiotics such as trimethoprim/sulfamethoxazole (Septra or Bactrim) may prevent relapses, especially in the nose.

Plasmapheresis, a method of removing ANCA antibodies from blood, may be performed in the hospital. It may help if a patient has bleeding in the lung or kidney failure.

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 (Prednisone, Solu-Medrol)

- 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 does decreases
- Weight gain – you may gain weight when you are on larger doses of steroids, especially in your face. Weight gain in your face is sometimes referred to as "moon 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, platelets, and red blood cells. This means you are at a higher risk for infection, and should try to stay away from crowds or people who are sick. Washing your hands regularly is recommended
- Nausea and vomiting – anti-nausea medicines can relieve your symptoms
- To help prevent damage to your bladder, try to drink 8-10 glasses of fluids a day.
- Your hair may thin as a result of taking the medicine, but will grow back after you finish.

Azathioprine (AZA)

- You may have a drop in your white blood cells, platelets, and red blood cells. This means you are at a higher risk for infection, and should try to stay away from crowds or people who are sick. Washing your hands regularly is recommended
- 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 (MMF)

- You may have a drop in your white blood cells, platelets, and red blood cells. This means you are at a higher risk for infection, and should try to stay away from crowds or people who are sick. Washing your hands regularly is recommended
- Nausea, vomiting, diarrhea. Taking the medicine in 3 divided doses rather than twice a day may decrease the nausea or vomiting (always consult your doctor before changing the way you take your medicine).

Rituximab:

- Some people may experience allergic side effects to the infusion. These may include low blood pressure, shortness of breath, and skin rashes.
- Other side effects related to the infusion include abnormal heart rhythm, nausea, and vomiting.

Septra/Bactrim:

- 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, platelets, and red blood cells. This means you are at a higher risk for infection, and should try to stay away from crowds or people who are sick. Washing your hands regularly is recommended.
- Nausea and vomiting – anti-nausea medicines can relieve your symptoms
- Mouth sores
- Bleeding from gums, nose
- Blood found in urine or bowel movement

What will happen after treatments are finished?

Following your first treatment phase, you may have a long period of remission. 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 any chronic illness. You may have tests to make sure your illness is a relapse of the vasculitis and not symptoms of 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.

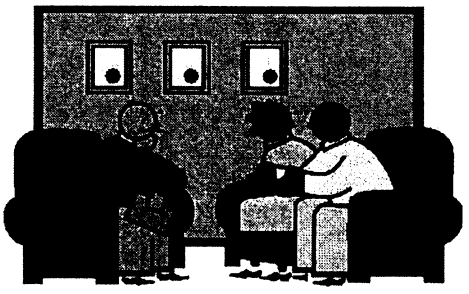
What is the outlook for patients with vasculitis?

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, a counselor or therapist or a minister can listen to your thoughts and concerns and help you find ways to cope.

The resources page in the back of this booklet has a listing of organizations that provide education and support to vasculitis patients. Also, ask your social worker, nurse or doctor if they of any local support groups in your area.



Resources

University of North Carolina Kidney Center (UNCKC) - The UNC Kidney Center's mission is to reduce the burden of chronic kidney disease through discovery about the pathophysiology and therapeutics of kidney disease and the development and assessment of educational programs about kidney disease for North Carolinians and their primary care physicians. The UNCKC maintains a vasculitis patient registry and their website contains useful links and general information for kidney disease patients, their practitioners, and their families. Internet: www.unckidneycenter.org. Printable copies of this brochure are available at this site, or by calling 1-866-462-9371 (toll-free).

The Vasculitis Foundation - (formerly the Wegener's Granulomatosis Association) a non-profit organization dedicated to providing emotional and informational support to patients with Wegener's granulomatosis (WG) and other types of vasculitis, to assisting them and their families in understanding the disease and recovery process, to educating and raising awareness in the general public and the medical community about WG and vasculitis, and to supporting research into the cause and cure of these diseases. For more information, please call 1(800)277-9474.
Internet:www.vasculitisfoundation.org

National Organization for Rare Diseases - a national group that serves as a clearinghouse for information about a number of rare diseases. Members receive a periodic newsletter and organize an annual national meeting. For more information, write NORD, P.O. Box 8923, Fairfield, CT 06812 or
Internet: http://www.rarediseases.org/

American Autoimmune Related Diseases Association - a national association that provides public outreach, and education for autoimmune diseases, including vasculitis, Wegener's Granulomatosis, and Polyarteritis Nodosa (PAN). This association publishes a quarterly newsletter and runs a referral line that provides information, support, and referrals. For more information, write 22100 Gratiot Ave., E. Detroit, MI 48021, or call (800) 598-4668. Internet: www.aarda.org

Polyarteritis Nodosa Research and Support Network (PAN Support) - this web-based network has a weekly chat room and an online discussion group for those afflicted with PAN and other autoimmune diseases. This website also provides educational materials and resources for PAN patients.
Internet: www.pansupport.org/.

Churg-Strauss Syndrome International Support Group - this group aims to provide information, encouragement and fellowship to Churg-Strauss Syndrome patients, care-givers, friends, physicians and relatives and to stimulate debate about the condition ensuring it receives wider recognition. This group maintains an online listserv for patient interaction and support. The North American (USA) Coordinator is Susan Guza: P.O. Box 328, Donald, OR, 97020-0328. E-mail: saguza@aol.com. Internet: www.churg-strauss.com.

Arthritis Foundation- a national group to support research to find the cure for and prevention of arthritis and to improve the quality of life for those affected by arthritis. For more information, write the national office at 1330 West Peachtree St., Atlanta, GA, 30309, or call (404) 872- 7100.
Internet: http://www.arthritis.org

National Kidney Foundation (NKF) - a national group that seeks to improve the care and treatment of those afflicted with diseases of the kidney and urinary tract through advances in disease detection, diagnosis and treatment. For more information, write National Kidney Foundation, 30 East 33rd Street, New York, New York 10016 or call 1-800-622-9010 or Internet: http://www.kidney.org

American Association of Kidney Patients (AAKP) - the American Association of Kidney Patients (AAKP) exists to serve the needs, interests and welfare of all kidney patients and their families. Its mission is to improve the lives of fellow kidney patients and their families by helping them to deal with the physical, emotional and social impact of kidney disease. To accomplish these goals, AAKP engages in a variety of educational and supportive programs, and publish several magazines and newsletters. They also have an annual meeting just for kidney patients. For more information write American Association of Kidney Patients, 3505 E. Frontage Rd., Ste. 315, Tampa, FL 33607, or call 1-800-749-2257, or go online at www.aakp.org.

Resources for 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.