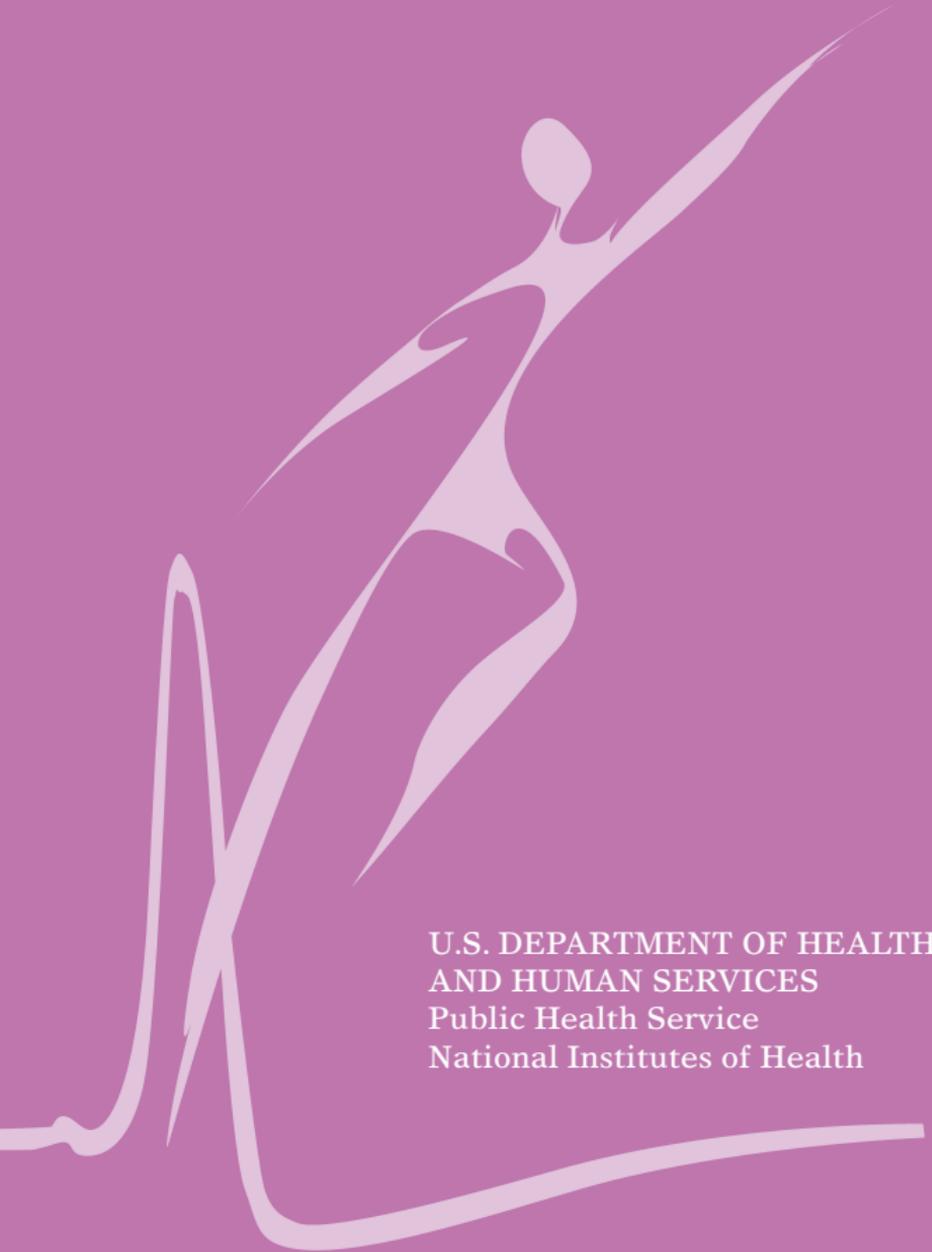


Vasculitis Syndromes of the Central and Peripheral Nervous Systems

A stylized white graphic on a purple background. It features a silhouette of a person with their arms raised in a gesture of triumph or hope. Below the figure is a white pulse line, similar to an ECG, that starts on the left and extends across the bottom of the page.

U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
Public Health Service
National Institutes of Health



Vasculitis Syndromes of the Central and Peripheral Nervous Systems

What is vasculitis?

Vasculitis is an inflammation of blood vessels, which includes the veins, arteries, and capillaries. Depending on the type, vasculitis can affect blood vessels of any type, size, or location. Inflammation occurs with infection or is thought to be due to a faulty immune system response. Dysfunction may occur due to the inflammation itself or over time as the blood vessel walls swell, harden, thicken, and develop scar tissue. This narrows the passage through which blood can flow. As the condition progresses, it can slow or completely stop the normal flow of blood.

How does vasculitis affect the nervous system?

Vasculitis can cause problems in any organ system, including the central (CNS) and peripheral (PNS) nervous systems. Vasculitic disorders, or syndromes, of the CNS and PNS are characterized by the presence of inflammatory cells in and around blood vessels, and secondary narrowing or blockage of the blood vessels that nourish the brain, spinal cord, or peripheral nerves. Any type or size of blood vessel may be involved—arteries, arterioles, veins, venules, or capillaries.

What are the symptoms?

A vasculitis syndrome may begin suddenly or develop over time. Symptoms include:

- headaches, especially a headache that doesn't go away
- fever
- malaise (feeling out-of-sorts)
- rapid weight loss
- confusion or forgetfulness leading to dementia
- aches and pains in the joints and muscles
- pain while chewing or swallowing
- paralysis or numbness, usually in the arms or legs
- visual disturbances, such as double vision, blurred vision, or blindness
- seizures, convulsions
- stroke or transient ischemic attack (TIA, sometimes also called a “mini-stroke”)
- unusual rashes or skin discoloration
- problems with the kidneys or other organs

How are these syndromes diagnosed?

A doctor who suspects CNS or PNS vasculitis will gather a comprehensive medical history of the individual, perform a physical examination, order laboratory tests (primarily blood tests), and recommend any other tests that seem appropriate. Electromyography and nerve conduction studies identify blocks and loss of nerve supply to muscle due to vasculitic nerve damage.

Diagnostic imaging of the brain blood vessels such as magnetic resonance or computed tomography angiograms can sometimes identify narrowing in the larger blood vessels. Direct injection of a contrast dye into brain blood vessels may be needed to look for narrowings consistent with vasculitis in medium-sized brain arteries.

However, the diagnosis of vasculitis often requires evidence that there is ongoing inflammation. Inflammatory cells may be found in the spinal fluid. Often there is a need to conduct a tissue biopsy to examine blood vessels under a microscope. In some cases a brain biopsy may be necessary to evaluate the compromised tissue. A definitive diagnosis is important because the treatment usually requires powerful immune-suppressive drugs. In addition, it is important to make sure that an infection is not causing the inflammation.

What are some of these syndromes called?

The diagnosis of a CNS or PNS vasculitis disorder will depend upon the number of blood vessels involved, their size, and their location in the CNS or PNS as well as the types of organs involved. Although these disorders are rare, there are many of them. Some of the better understood syndromes are:

Temporal arteritis (also called giant cell arteritis or cranial arteritis)

Temporal arteritis is a common chronic inflammatory disease of large blood vessels occurring primarily in people 50 and older. It most often involves narrowing and sometimes blockage of the arteries that bring blood to

the brain. Doctors will diagnose temporal arteritis if at least three of the following symptoms are present:

- new, severe headache
- visual disturbances
- pain in the jaw or tongue when chewing or swallowing
- tenderness in the temporal arteries (the arteries that run across the temples on either side of the head) or the scalp

Fever, weight loss, and neck or muscle pain can occur, usually in the early phase of the disease. Individuals may also have arthritis; carpal tunnel syndrome; fatigue; and weakness, paralysis, or numbness in isolated muscles. The disease is usually limited to one to two years and is rarely fatal.

Abrupt but reversible blindness is the most dramatic complication of temporal arteritis. About one in ten individuals with temporal arteritis will develop blindness in one eye, preceded by visual disturbances. Once one eye is affected, three out of four individuals will go on to lose vision in the other eye, most in two weeks or less.

The main goal of treatment for temporal arteritis is to prevent blindness. Most individuals respond well to steroid drugs, such as prednisone and methylprednisolone, but they must be given promptly and carefully monitored. Long-term use of steroids can cause harmful side effects, such as collapsing vertebrae, muscle pain, diabetes, cataracts, and infection.

Primary angiitis of the CNS (granulomatous angiitis)

The symptoms of this rare disorder develop slowly. Symptoms include headache and encephalopathy-like symptoms such as dementia and tremor. Stroke, TIA, and seizures can occur. Definitive diagnosis may require brain biopsy. Treatment includes steroid and immunosuppressive drugs, such as prednisolone and cyclophosphamide. It is fatal if left untreated.

Takayasu's disease

This disease affects large arteries such as the aorta, which brings blood to the arms, legs, and head. It primarily strikes individuals of Asian descent and predominantly affects females under the age of 40. The main symptoms are fainting and visual disturbances and it may also cause stroke. Although the disorder is serious, the prognosis is positive: more than 90 percent of those diagnosed with Takayasu's disease survive beyond a decade after diagnosis. Steroid drugs are used in the early phase of the disease, but some individuals become steroid-resistant and have to switch to cyclophosphamide or low-dose methotrexate.

Periarteritis nodosa

The onset of this rare and serious disease is generally between the ages of 40 and 50, but it can occur at any age. Men are three times more likely to develop the disease than women.

Symptoms can mimic those of many other diseases, but the most common initial complaints are fever, abdominal pain, numbness or pain in the legs and limbs, weakness, and

unexplained weight loss. As the disease progresses, the kidneys may fail and high blood pressure may develop rapidly. Certain drugs (for example, those in the sulfa family), vaccines, bacterial infections, and viral infections have been associated with the onset of the disease. Damage to the PNS with neuropathy is more common than damage to the CNS, but if the disease does involve the CNS, damage to brain and spinal cord tissue can occur.

The disease is treated aggressively with high doses of steroids and immunosuppressive drugs such as cyclophosphamide. Eighty percent of individuals who receive appropriate treatment are alive five years later. Untreated disease is often fatal, ending in heart failure, kidney failure, or failure of other vital organs.

Are there additional vasculitis disorders that can cause neurological symptoms?

Other vasculitis syndromes include Kawasaki disease, which can cause stroke or encephalopathy in children; Churg-Strauss syndrome; Wegener's granulomatosis; systemic lupus erythematosus; scleroderma; rheumatoid arthritis; Sjogren's syndrome; and Behcet's disease.

What research is being done to better understand these syndromes?

The National Institute of Neurological Disorders and Stroke (NINDS), a component of the National Institutes of Health (NIH), and other NIH institutes conduct research relating to vasculitis syndromes in laboratories at the NIH and also support vasculitis

research through grants to major medical institutions across the country.

The NINDS supports The Vasculitis Clinical Research Consortium (VCRC), a network of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research and improving the care of individuals with vasculitis, including Wegener's granulomatosis, microscopic polyangiitis, Churg-Strauss syndrome, polyarteritis nodosa, Takayasu's arteritis, and temporal arteritis. The medical centers are located at Boston University School of Medicine, Cleveland Clinic Foundation, The Johns Hopkins Vasculitis Center, and Mayo Clinic College of Medicine. The Consortium's internet site provides information about clinical research and clinical trial opportunities and helps individuals connect with expert doctors and patient support groups.

Where can I get more information?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN

P.O. Box 5801

Bethesda, MD 20824

800-352-9424

www.ninds.nih.gov

Information also is available from the following organizations:

American Autoimmune Related Diseases Association

22100 Gratiot Avenue
East Detroit, MI 48021-2227
586-776-3900
800-598-4668
www.aarda.org

National Organization for Rare Disorders (NORD)

P.O. Box 1968
(55 Kenosia Avenue)
Danbury, CT 06813-1968
203-744-0100 Voice Mail
800-999-NORD (6673)
www.rarediseases.org

National Eye Institute (NEI)

National Institutes of Health, DHHS
31 Center Drive, Rm. 6A32 MSC 2510
Bethesda, MD 20892-2510
301-496-5248
www.nei.nih.gov

National Institute of Allergy and Infectious Diseases (NIAID)

National Institutes of Health, DHHS
6610 Rockledge Drive, MSC 6612
Bethesda, MD 20892-6612
301-496-5717
www.niaid.nih.gov





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