

CENTRAL NERVOUS SYSTEM VASCULITIS

What is central nervous system (CNS) vasculitis?

Central nervous system (CNS) vasculitis is among a family of rare disorders characterized by inflammation of the blood vessels, which restricts blood flow and damages vital organs and tissues. A serious condition, CNS vasculitis can block the vessels that supply the brain and spinal cord, causing potentially life-threatening complications such as loss of brain function, or stroke.

CNS vasculitis is typically categorized as “primary” and “secondary”:

- Primary angiitis of the CNS (PACNS) is vasculitis confined specifically to the brain and spinal cord, which make up the central nervous system. It is not associated with any other systemic (affecting the whole body) disease.
- Secondary CNS vasculitis usually occurs in the presence of other autoimmune diseases such as systemic lupus erythematosus, dermatomyositis, or rheumatoid arthritis; systemic forms of vasculitis, such as granulomatosis with polyangiitis, microscopic polyangiitis, or Behcet’s syndrome; or viral or bacterial infections.

Diagnosis of this condition can be challenging because a number of other diseases and infections have similar symptoms. Once diagnosed, CNS vasculitis is typically treated with corticosteroids such as prednisone, used in combination with medications that suppress the immune system. Even with effective treatment, relapse of CNS vasculitis is common, so ongoing medical follow-up is important.

Causes

The cause of CNS vasculitis is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder—a disease which occurs when the body’s natural defense system mistakenly attacks healthy tissues. Researchers believe an infection may contribute to the onset of CNS vasculitis. Environmental and genetic factors may play a role as well.

Who gets vasculitis of the CNS?

In general, CNS vasculitis is considered rare. In the case of PACNS, the disorder can affect people of all ages but generally peaks around age 50. It most often occurs in males.

Symptoms

Many forms of vasculitis are accompanied by fever, fatigue, sudden weight loss, or skin rashes. When vasculitis affects the central nervous system, symptoms may include:

- Severe headaches that don’t go away
- Strokes or transient ischemic attacks (mini-strokes)
- Swelling of the brain (encephalopathy)
- Forgetfulness or confusion
- Muscle weakness or paralysis
- Difficulty with coordination
- Abnormal sensations or loss of sensations

CENTRAL NERVOUS SYSTEM VASCULITIS

- Vision problems
- Seizures or convulsions

Diagnosis

Diagnosing CNS vasculitis poses a challenge for physicians. Many of the key symptoms of CNS vasculitis are shared by other diseases and infections, so these “mimics” must be ruled out. There is no single diagnostic test for CNS vasculitis, so your doctor will consider a number of factors, including a detailed medical history, a physical examination, laboratory tests, and specialized imaging studies. A biopsy of tissue from blood vessels in the brain or spine is usually required to confirm a diagnosis.

If CNS vasculitis is suspected, your doctor will likely order the following tests:

- **Lab work:** Blood tests are frequently normal in PACNS vasculitis, but may be abnormal if reflecting another underlying disease.
- **Examination of the spinal fluid:** A sample of the cerebrospinal fluid (which surrounds the brain) is removed through a spinal tap and analyzed for infection and signs of inflammation.
- **Diagnostic imaging:** Computed tomography (CT) scans and magnetic resonance imaging (MRI) produce images that can help identify abnormalities of the brain, spinal cord, blood vessels, and other organs and tissues.
- **Cerebral angiogram:** An angiogram detects blockages of blood vessels using X-rays taken during the injection of a contrast agent.
- **Biopsy:** This surgical procedure removes a small tissue sample from a blood vessel or an affected organ, which is examined under a microscope for signs of inflammation or tissue damage. Because other conditions can cause similar brain vessel abnormalities as CNS vasculitis, a brain biopsy may be the only way to make a definitive diagnosis.

A major mimic of CNS vasculitis

Reversible cerebral vasoconstriction syndrome (RCVS) refers to a group of conditions that involve spasm of the brain vessels, and that mimic CNS vasculitis. RCVS features sudden, severe headaches, as well as strokes or bleeding into the brain. It is essential for clinicians evaluating patients to be aware of RCVS and to distinguish it from CNS vasculitis, given the differences between the two diseases, including the treatment and prognosis.

Treatment

CNS vasculitis is typically treated with a high-dose corticosteroid, such as prednisone, to reduce inflammation. For more severe cases, prednisone is used in combination with drugs that suppress the immune system’s response, such as cyclophosphamide, mycophenolate mofetil or azathioprine. Treatment may be aggressive for the first six months and then tapered down as symptoms improve.

In addition to medication, other forms of treatment may include physical, occupational or speech therapy. If memory is affected, brain activities that enhance memory may be recommended.

CENTRAL NERVOUS SYSTEM VASCULITIS

Side effects

The medications used to treat CNS vasculitis have potentially serious side effects, such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it's important to see your doctor for regular checkups. Medications may be prescribed to offset side effects. Infection prevention is also important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment, relapses are common for individuals with CNS vasculitis. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular check-ups and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your medical team

Effective treatment of CNS vasculitis may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, you may need to see the following specialists: rheumatologist (joints, muscles and immune system); neurologist (brain/nervous system); physical, occupational or speech therapist; or others as needed.

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place. To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it's up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

Living with CNS vasculitis

Living with a chronic disease such as CNS vasculitis can be overwhelming at times. Fatigue, pain, emotional stress and medication side effects can take a toll on your sense of well-being, affecting relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

There is no cure for CNS vasculitis at this time, however it is treatable. Early diagnosis and treatment are essential to prevent potentially life-threatening loss of brain function or stroke. Other diseases often have the same symptoms as CNS vasculitis, so accurate diagnosis involves ruling out these conditions. Even with treatment, relapses are common with CNS vasculitis, so follow-up medical care is essential.

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients

CENTRAL NERVOUS SYSTEM VASCULITIS

to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information. For more information on vasculitis research, visit: www.vasculitisfoundation.org/research

About Vasculitis

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

A Family of Diseases

- Anti-GBM (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

About the Vasculitis Foundation

The Vasculitis Foundation (VF) is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services. To learn more, and get the most updated disease and treatment information, visit www.vasculitisfoundation.org

Connect with the VF on Social Media

- Instagram: [vasculitisfoundation](https://www.instagram.com/vasculitisfoundation)
- Twitter: [@VasculitisFound](https://twitter.com/VasculitisFound)
- VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation

CENTRAL NERVOUS SYSTEM VASCULITIS

Ways to Get Involved

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium
- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

Vasculitis Foundation Mission

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

Join the VPPRN!

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: www.VPPRN.org

The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: www.rarediseasesnetwork.org/cms/vcrc

Vasculitis Foundation

PO Box 28660
Kansas City, Missouri 64188-8660
USA
Phone: 816.436.8211
Toll-free: 800.277.9474
Fax: 816.656.3838
www.VasculitisFoundation.org
Email: vf@vasculitisfoundation.org

The VF gratefully acknowledges Rula Hajj-Ali, MD, Cleveland Clinic, for her expertise and contribution to this brochure. This brochure was supported by an unrestricted education grant from Genentech. The Vasculitis Foundation is solely responsible for all content.

The Vasculitis Foundation (VF) is a 501 (c) (3) nonprofit organization governed by a Board of Directors and advised on medical issues by a Medical and Scientific Advisory Board. VF's educational materials are not intended to replace the counsel of a physician. VF does not endorse any medications, products or treatments for vasculitis, and advises you to consult a physician before initiating any treatment.

(Updated October 2018)