

CUTANEOUS SMALL-VESSEL VASCULITIS (formerly hypersensitivity vasculitis)

What is cutaneous small-vessel vasculitis (CSVV)?

Cutaneous small-vessel vasculitis (CSVV) is among a family of rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Formerly called hypersensitivity vasculitis, this disorder most commonly affects the skin. It typically occurs in individuals 16 or older. CSVV is also known as leukocytoclastic vasculitis and allergic vasculitis.

Common symptoms of CSVV include a purple or reddish rash over the legs, buttocks or torso, and sometimes the upper body. Hives, blisters, and open sores can also occur. When CSVV is systemic (affecting the whole body), it typically affects the joints, gastrointestinal tract and kidneys. In children, the disease can resemble IgA vasculitis (formerly called Henoch-Schönlein purpura), which also affects the small vessels of the skin and causes a similar rash.

CSVV is often triggered by an allergic reaction to a drug or by an infection, and usually clears up once the offending medication is discontinued or the infection is treated. However, some patients will need corticosteroids used in combination with medications that suppress the immune system, to control inflammation. If there is no organ involvement, most cases go away within weeks to months. However, CSVV can be chronic, with periods of relapse and remission, so ongoing medical care may be necessary.

Causes

The exact cause of CSVV 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 tissue. CSVV can be caused by an allergic reaction to drugs, such as antibiotics, diuretics and blood pressure medicines, as well as foods or food additives. Other triggers include upper respiratory tract infections, and viruses such as hepatitis B or C, and HIV. Cancer is a rare but possible cause of CSVV.

CSVV can affect individuals with other autoimmune diseases such as rheumatoid arthritis, Crohn’s disease, systemic lupus erythematosus and Sjögren’s syndrome, as well as certain forms of larger-vessel vasculitis.

Who gets CSVV?

CSVV can occur at any age, but usually affects individuals over the age of 16. The disease occurs in females and males equally, although some studies indicate it is slightly more common in men. It most often affects Caucasians.

An estimated 38 to 55 per million adults per year are diagnosed with CSVV annually. A 2014 population-based study in Minnesota found the incidence of CSVV in adults to be 45 cases per million. According to studies, CSVV has a reported incidence in Spain of 10 to 30 cases per million people per year.

Symptoms

The hallmark symptom of CSVV is a skin rash, with tender, purple or reddish-brown spots covering large areas—especially the legs, buttocks or torso—or upper body. Other symptoms include blisters, hives and open sores (ulcers) with dead tissue. Crops of lesions typically appear over one or two days, with

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more appearing over the next days to weeks, until treatment is initiated or the allergy or other trigger goes away. Individual lesions can last up to three weeks and leave scarring, especially if they ulcerate.

Diagnosis

There is no single test to diagnose CSVV. Your doctor will likely take a detailed drug history and ask about current medications you've been taking, as well as recent infections. A typical exam includes a thorough medical history; a physical examination; blood and urine tests; and specialized imaging studies such as X-rays and computed tomography (CT) scans, when indicated.

A skin biopsy is typically ordered. Biopsy is a surgical procedure that removes a small sample of affected tissue, which is examined under a microscope for signs of inflammation or damage. Usually a biopsy of the skin rash that shows inflammation of the small blood vessels confirms the diagnosis of CSVV.

Treatment

Treatment of CSVV starts with addressing the suspected trigger. If a drug may have caused the disease, your doctor will probably advise you to discontinue it. If the medication is the culprit, your symptoms should disappear within a few weeks. If an infection is the suspected cause, treating it may resolve symptoms. If the trigger was a food allergy, removing the offending food from the diet may help.

- Because the legs are commonly affected by rash or joint pain, avoidance of prolonged standing can be helpful, along with elevation of the legs and use of compression stockings.
- To reduce inflammation in the blood vessels, your doctor may prescribe nonsteroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen or naproxen, or corticosteroids.
- In patients with more severe or ongoing skin rashes that are not due to infection, drugs such as colchicine, antihistamines, hydroxychloroquine, and dapsone (or a combination of these drugs) may be helpful to control symptoms.
- In severe disease, a high-dose steroid may be required. In addition, immunosuppressive drugs such as azathioprine and methotrexate may be prescribed.
- Patients with disease in organs beyond the skin should be referred to a specialty doctor.

Side effects

The medications used to treat CSVV 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 very 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, some individuals with CSVV will experience relapses. If your 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.

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Your medical team

Effective treatment of CSVV 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, immune system); dermatologist (skin); 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 CSVV

Living with a chronic condition such as CSVV 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

In general, CSVV patients who primarily have skin and or/joint symptoms have a good outlook, with the disease typically resolving within a few weeks or months. Some develop chronic, recurring disease, which can affect quality of life, and in those cases 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 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

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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

Ways to Get Involved

- Participate in research
- Join social media

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- 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

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