



## **What is Cutaneous Small-Vessel Vasculitis?**

Cutaneous small-vessel vasculitis (CSVV) is a form of vasculitis that affects the skin. CSVV is also known as leukocytoclastic vasculitis and allergic vasculitis, and was formerly called hypersensitivity vasculitis.

Vasculitis is a group of rare disorders that are associated with inflammation of the blood vessels. Inflammation of blood vessels may result in narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues.

Vasculitis is classified as an autoimmune disorder—a disease that occurs when the body’s natural defense system mistakenly attacks healthy tissue.

## **Causes**

The exact cause of CSVV is not fully understood. It 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 causes include:

- Infections
- Viruses such as hepatitis B or C, and HIV
- Cancer – a rare but possible cause of CSVV

## **Who Gets CSVV?**

CSVV can occur at any age. The condition occurs in females and males equally, although some studies indicate it is slightly more common in men. It most often affects Caucasians.

CSVV can affect individuals with other autoimmune diseases such as rheumatoid arthritis, Crohn’s disease, systemic lupus erythematosus and Sjögren’s syndrome.

## Symptoms

Common symptoms of CSVV include:

- A skin rash with tender, purple or reddish-brown spots covering large areas —especially the legs, buttocks or torso—or upper body
- Blisters, hives and open sores (ulcers) can occur

A cluster of skin lesions typically appear over one or two days, with more appearing over the next days to weeks.

The symptoms may go away after treatment is started, or the allergy or other triggers goes away. Individual skin lesions can last up to three weeks or longer and can leave scarring, especially if they ulcerate.

Vasculitis affecting the skin (cutaneous) can be a single organ vasculitis (when limited to the skin), such as seen in CSVV, or it can be part of a systemic (whole body) vasculitic disorder (e.g., GPA, Ig-A vasculitis and other small vessel vasculitides).

## Diagnosis

The diagnosis of CSVV includes multiple tools:

- A thorough medical history
- A physical examination
- Blood and urine tests
- Specialized imaging studies such as x-rays and computed tomography scans, when indicated
- A detailed drug history to determine current medications and recent infections
- Skin biopsy

A skin biopsy is typically ordered (a 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 cause:

- 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 may disappear within a few weeks of stopping the medication.
- If an infection is the suspected cause, treating it may resolve symptoms.
- If the cause was a food allergy, removing the offending food from the diet may help.
- Because the legs are commonly affected by a 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, naproxen, or corticosteroids.

In patients with more severe or recurrent skin rashes without an obvious cause, other medications may be helpful to control symptoms, such as:

- Colchicine, antihistamines, hydroxychloroquine, and dapsone (or a combination of these drugs)

In severe disease, a high-dose steroid may be required. In addition, immunosuppressive drugs such as azathioprine and methotrexate may be prescribed.

## Side Effects of the Medications

The medications used to treat CSVV have potentially serious side effects, such as:

- Lowering your body's ability to fight infection
- 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.

## **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 and 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. This can affect 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.

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