

URTICARIAL VASCULITIS

What is urticarial vasculitis?

Urticarial vasculitis 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. This form of vasculitis primarily affects the small vessels of the skin, causing red patches and hives that can itch, burn and leave skin discoloration. Depending on the form of urticarial vasculitis, other organ systems may be affected.

There are two categories of urticarial vasculitis named for the level of “complement proteins” in the blood, which play a role in the immune system.

- Normocomplementemic urticarial vasculitis refers to a normal level of complement proteins and is usually less severe, having little if any systemic (affecting multiple organs) involvement.
- Hypocomplementemic urticarial vasculitis refers to low levels of complement proteins and is more severe, having systemic involvement typically affecting the joints, lungs, kidneys, gastrointestinal tract and eyes.

Treatment depends on the extent of symptoms and organ involvement. When the disease primarily affects the skin, antihistamines or anti-inflammatory drugs such as ibuprofen or naproxen may relieve symptoms. For more severe cases, corticosteroids such as prednisone and/or other powerful drugs that suppress the immune system may be prescribed. Urticarial vasculitis can be a difficult-to-treat, chronic illness that can cause serious health problems, so ongoing medical care is essential.

Causes

The cause of urticarial 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 tissue. In urticarial vasculitis, the inflammatory process may be set in motion by an infection or virus such as hepatitis, a drug reaction, or the existence of cancer or another autoimmune disorder such as systemic lupus erythematosus, rheumatoid arthritis or Sjögren’s syndrome.

Who gets urticarial vasculitis?

Urticarial vasculitis can affect people of any age, but it most commonly occurs in adults between 30 to 40 years of age. Women get the disease about twice as often as men. Urticarial vasculitis is considered rare, however its precise frequency in the United States and worldwide is unknown.

Symptoms

Urticarial vasculitis usually begins with an eruption of skin lesions (wheals) and hives (urticaria), which cause itching, pain and burning sensations. Skin patches are often red-rimmed with white centers, and may have petechia—red or purple pinpoint spots caused by bleeding under the skin. The patches can be present for more than 24 hours and may leave a bruise-like skin discoloration as they heal.

Those with more severe, systemic involvement may also experience:

- Fever
- Joint pain
- Swelling of tissues under the skin
- Abdominal pain
- Swollen lymph glands

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- Shortness of breath
- Bloodshot or inflamed eyes
- Kidney inflammation
- Cardiac involvement

Complications

Common complications of urticarial vasculitis include skin pigmentation and occasionally skin ulcers. In more serious cases, individuals may have damage to the lungs and suffer chronic obstructive pulmonary disease, as well as eye and kidney complications.

Diagnosis

There is no single diagnostic test for 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 skin biopsy—surgical removal and analysis of a small tissue sample—can show inflammation and damage to the small blood vessels, helping to confirm the diagnosis. Low blood levels of certain complement proteins help establish the diagnosis of hypocomplementemic urticarial vasculitis.

Because urticarial vasculitis is often associated with other diseases, it may be necessary to do other tests to rule out underlying conditions. Tests of vital organs, such as the kidneys and lungs, may also be indicated, especially when the blood levels of complement are low.

Treatment

The course of treatment for urticarial vasculitis depends on the extent of symptoms and organs affected. When symptoms primarily affect the skin, lesions may resolve on their own or may be relieved with minimal treatment such as antihistamines and NSAIDs such as ibuprofen or naproxen.

In cases of more severe organ involvement, drugs that affect the immune system may be required, such as prednisone, hydroxychloroquine, colchicine, or dapsone; or immunosuppressive agents such as azathioprine, methotrexate or cyclophosphamide. Treatment may be intermittent for some, but many patients need to take medications for several years.

Side effects

The medications used to treat urticarial 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 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, urticarial vasculitis can be a chronic disease with periods of relapse and remission. 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 are important in detecting relapses early.

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Your medical team

Effective treatment of urticarial vasculitis may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, patients with urticarial vasculitis may need to see the following: dermatologist (skin); rheumatologist (joints, muscles, immune system); pulmonologist (lungs); nephrologist (kidneys); immunologist (allergies); 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. Urticarial vasculitis can be difficult to treat, so a second opinion from a specialist doctor is highly recommended.

Living with urticarial vasculitis

Living with a chronic condition such as urticarial 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 urticarial vasculitis at this time. For most people, urticarial vasculitis is a disease of the skin, with a minority of patients developing systemic organ involvement. When it is related to an existing autoimmune disease such as lupus, or cancer, the prognosis may depend on the underlying disease. Because urticarial vasculitis may become chronic, 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

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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

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