

## **What is Urticarial Vasculitis?**

Urticarial vasculitis is a form of vasculitis, which is an 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.

Urticarial vasculitis primarily affects the small vessels of the skin, causing red patches and hives that can itch, burn and leave the skin discolored. 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 that 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 usually more severe, having systemic involvement typically affecting the joints, lungs, kidneys, gastrointestinal tract, and eyes.

## **Causes**

The cause of urticarial vasculitis is not fully understood. It is classified as an autoimmune disorder, which is a disease that 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
- The existence of cancer
- Another autoimmune disorder such as systemic lupus erythematosus, rheumatoid arthritis, or Sjögren’s syndrome

## Who Gets Urticarial Vasculitis?

Urticarial vasculitis is considered rare but can affect people of any age, and most commonly in adults between 30 to 40 years of age.

## Symptoms

Urticarial vasculitis usually begins with 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
- Shortness of breath
- Bloodshot or inflamed eyes
- Kidney inflammation
- Cardiac involvement

## Complications

Common complications of urticarial vasculitis include skin discoloration 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 such as a detailed medical history and physical examination, as well as:

- Laboratory tests such as complete blood cell count, sedimentation rate, c-reactive protein, and complement proteins (low blood levels of certain complement proteins help establish the diagnosis of hypocomplementemic urticarial vasculitis).
- Other tests may be necessary, such as pulmonary functions tests and chest computed tomography.

- 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.

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 be relieved with minimal treatment such as antihistamines and nonsteroidal anti-inflammatories 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
- 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.

## 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)
- 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.

A health care journal is helpful to track medications, symptoms, test results, and notes from doctor appointments.

Make a list of questions before your doctor visit. 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. 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

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.

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