CONDITION

Necrotizing vasculitis

Necrotizing vasculitis is severe blood vessel inflammation that damages the vessel wall and may reduce blood flow to organs.

WHAT IT IS
Destructive inflammation of blood vessel walls

BODY SYSTEM
Blood vessels, potentially affecting many organs

MAIN SYMPTOMS
Fatigue, fever, skin changes, organ-specific problems

WHAT CAUSES IT
Immune disorders, infection, medicines, or unknown

HOW IT IS DIAGNOSED
Tissue biopsy, supported by blood, urine, and imaging tests

MAIN TREATMENT
Cause-directed therapy, often immune-suppressing medicines

CAN IT BE CURED?
Sometimes - cause and organ damage matter

IS IT AN EMERGENCY?
Sometimes - breathing trouble, coughing blood, or stroke signs

LONG-TERM OUTLOOK
Often manageable, but relapses and permanent damage can occur

Overview

Necrotizing vasculitis is a pattern of severe inflammation and damage in blood vessel walls. "Necrotizing" means that part of the vessel wall has been injured or destroyed. "Vasculitis" means inflammation of blood vessels.

It is a condition rather than one specific disease. Several diseases can produce this type of blood vessel damage. Some mainly affect small vessels, while others affect medium-sized or larger arteries.

Inflamed vessels may become narrowed, blocked, weakened, or leaky. This can reduce the supply of oxygen-rich blood to nearby tissue. The effects depend on which vessels and organs are involved. The skin may develop spots or sores, while inflammation involving the kidneys, lungs, nerves, brain, heart, or digestive tract can be more serious.

A tissue biopsy often provides the clearest evidence of necrotizing vasculitis. A biopsy is a procedure in which a small tissue sample is removed and examined under a microscope. Further testing is needed to identify the underlying type and guide treatment.

Symptoms

Symptoms can begin over days or weeks, although some forms develop more slowly. Early symptoms may be general and can occur with many other conditions. These include:

Other symptoms reflect the organ affected.

Skin vasculitis may cause raised purple or red spots, often on the lower legs. These spots are called purpura and do not fade when pressed. Reduced blood flow can also cause painful ulcers or darkened areas of skin.

Nerve inflammation may cause burning pain, numbness, tingling, or weakness. Symptoms can be uneven, such as sudden weakness in one foot or hand.

Kidney involvement often causes no pain. Possible signs include blood or protein in the urine, swelling around the ankles, higher blood pressure, or reduced kidney function. These changes may first appear on urine and blood tests.

Lung involvement can cause breathlessness, chest discomfort, cough, or coughing up blood. Nose and sinus inflammation may cause repeated sinus problems, crusting, nosebleeds, or hearing changes in some forms of vasculitis.

Reduced blood flow to the digestive tract can cause persistent or severe abdominal pain, nausea, vomiting, or blood in the stool. Brain or heart involvement may cause stroke-like symptoms, chest pain, or other urgent problems.

Having one of these symptoms does not mean that you have necrotizing vasculitis. Many are more commonly caused by other conditions.

- Tiredness, fever, night sweats, or unintended weight loss
- Muscle or joint aches
- Loss of appetite or a general feeling of being unwell
- Skin spots, painful lumps, blisters, ulcers, or purple discoloration

Causes and risk factors

Necrotizing vasculitis is often caused by an immune system problem. The immune system

normally protects the body from infection. In vasculitis, immune activity mistakenly damages blood vessels.

It can occur as part of several named conditions. Examples include granulomatosis with polyangiitis, microscopic polyangiitis, and polyarteritis nodosa. Some forms are linked to immune proteins called antibodies. ANCA, or antineutrophil cytoplasmic antibodies, are one example. ANCA may support a diagnosis, but a positive result alone does not prove that vasculitis is present.

Other cases are associated with infections, including certain hepatitis viruses. Less often, a medicine may trigger vessel inflammation. Vasculitis can also occur alongside another immune condition, such as lupus or rheumatoid arthritis, or occasionally with a blood disorder or cancer.

Sometimes no underlying trigger is found. This is called idiopathic vasculitis.

Risk is shaped mainly by the specific type of vasculitis and its cause. It is not usually the result of diet, stress, exercise habits, or something you did wrong. Most forms are not directly inherited, although genes may influence immune system risk.

Diagnosis

Diagnosis usually involves confirming blood vessel inflammation and working out its cause, extent, and severity. There is no single blood test that can diagnose every form.

Your assessment may include questions about skin changes, infections, new medicines, breathing, urine, nerve symptoms, and other health conditions. A physical examination looks for signs of reduced blood flow or organ inflammation.

Common investigations include:

Under a microscope, necrotizing vasculitis shows destructive inflammation within a vessel wall. Immune staining may help distinguish antibody-related disease from inflammation caused by immune-protein deposits.

A biopsy is not always possible or safe. Imaging of the arteries may instead show narrowing, blockage, or small bulges called aneurysms, particularly when medium-sized arteries are involved.

Results must be interpreted together. Infection, blood clots, cholesterol deposits, medicine reactions, and other conditions can sometimes resemble vasculitis.

- Blood tests for inflammation, blood cell counts, kidney and liver function, and immune antibodies

- Urine tests for blood and protein, which may reveal kidney involvement
- Tests for infections that can cause or resemble vasculitis
- Imaging such as ultrasound, CT, MRI, or blood vessel imaging
- A biopsy of affected skin, kidney, nerve, muscle, lung, or another accessible tissue

Treatment and management

Treatment is based on the underlying cause, the vessels involved, and whether important organs are at risk. Care may involve rheumatology, kidney, lung, skin, nerve, infectious disease, or other specialists.

When harmful immune activity is the cause, treatment usually uses medicines that reduce inflammation and suppress the immune response. Corticosteroids like prednisone or hydrocortisone are commonly used to bring inflammation under control. Depending on the specific diagnosis and severity, another immune-suppressing medicine or a targeted antibody treatment may be added.

Severe disease is often treated in two stages. The first aims to stop active vessel damage and achieve remission, meaning the inflammation is controlled. The second uses maintenance treatment to reduce the chance of relapse. The exact medicines and length of treatment differ among types of vasculitis.

If an infection is responsible, treating that infection is central. Immune suppression may be unsafe if an untreated infection is mistaken for immune-mediated vasculitis. If a medicine appears to be the trigger, the clinical team may advise stopping or replacing it. Do not stop a prescribed medicine without advice unless you are receiving emergency care for a suspected severe reaction.

Some people also need treatment for complications, such as kidney support, blood pressure management, wound care, pain rehabilitation, or physical therapy. Plasma exchange, which removes and replaces part of the liquid portion of blood, is reserved for selected situations rather than used routinely for everyone.

Monitoring is an important part of care. It may include blood pressure checks, urine tests, blood counts, kidney and liver tests, symptom reviews, and checks for treatment side effects. Medicines that suppress immunity can increase infection risk, so vaccination and infection-prevention plans may need review.

Complications

Complications result from reduced blood flow, bleeding from damaged vessels, inflammation within organs, or treatment side effects.

Possible complications include:

Not everyone develops these problems. Early recognition and effective control of inflammation can reduce the risk of further damage, although tissue already badly injured may not fully recover.

- Permanent kidney damage or kidney failure
- Lung bleeding or lasting breathing problems
- Nerve damage, chronic pain, weakness, or difficulty walking
- Skin ulcers, scarring, or tissue loss
- Bowel injury from reduced blood flow
- Stroke, heart injury, or weakened arteries
- Serious infection related to immune-suppressing treatment

Prevention

Most immune-mediated forms of necrotizing vasculitis cannot be reliably prevented. They are not usually caused by everyday choices.

Some secondary cases may be preventable. Vaccination against hepatitis B can prevent an infection associated with some cases of polyarteritis nodosa. Prompt treatment of relevant infections may also reduce complications. Avoiding a medicine that previously caused vasculitis may help prevent another reaction.

If you take immune-suppressing treatment, reducing infection exposure and keeping recommended vaccinations up to date can support safer care. Live vaccines may not be suitable during some treatments, so vaccination plans should be checked with your healthcare team.

Smoking does not usually cause necrotizing vasculitis, but it damages blood vessels and can add to heart, lung, and circulation risks. Stopping smoking may support overall vascular health.

Living with it and what to expect

The course differs according to the underlying type, the organs affected, how quickly treatment begins, and how well inflammation responds. Some infection- or medicine-related cases settle after the trigger is treated or removed. Immune-mediated forms may behave as long-term illnesses.

Treatment can bring many people into remission. Remission does not always mean that the condition is permanently cured. Some people remain well for years, while others have relapses that require further treatment.

A relapse means that active vessel inflammation has returned. Its symptoms may resemble the first episode, but they can also be different. Keep track of new skin changes, breathing problems, dark or bloody urine, worsening numbness, and unexplained general symptoms. Routine test results may sometimes reveal activity before you feel seriously unwell.

Fatigue can continue after inflammation is controlled. Recovery from nerve, kidney, lung, or muscle injury may take months. Some damage can be permanent, but rehabilitation, pain support, and practical adjustments may improve function.

Regular follow-up is usually needed even during remission. Visits help monitor organ function, medicine side effects, infection risk, and possible relapse. Do not change immune-suppressing treatment suddenly unless your clinical team tells you to do so.

Many people continue working, studying, exercising, travelling, and caring for their families. Activities may need to be adjusted during active illness or recovery. The long-term plan should reflect your symptoms, treatment effects, and any lasting organ damage.

When to get medical help

Arrange a prompt medical assessment if you develop unexplained purple skin spots, persistent fever, new numbness or weakness, blood in your urine, worsening swelling, repeated nosebleeds, or symptoms that could suggest inflammation in an organ. These symptoms need evaluation, but they are not always caused by vasculitis.

If you already have a vasculitis diagnosis, contact your healthcare team promptly about possible relapse symptoms or signs of infection. Fever during immune-suppressing treatment deserves timely advice because the treatment can make infections harder to fight.

Seek emergency care for:

Routine follow-up is appropriate for stable symptoms, planned test monitoring, medication reviews, and rehabilitation needs. Emergency symptoms should not wait for a routine appointment.

- Severe trouble breathing or coughing up blood
- New facial drooping, speech difficulty, confusion, or one-sided weakness
- Sudden severe chest or abdominal pain
- Fainting, severe weakness, or rapidly worsening illness
- A cold, pale, blue, very painful, or suddenly numb limb

Common questions

Is necrotizing vasculitis cancer?

No. It is inflammation that damages blood vessel walls, not uncontrolled growth of cancer cells. Rarely, vasculitis can occur alongside a cancer or blood disorder, so tests may look for an underlying condition when appropriate.

Is necrotizing vasculitis contagious?

The vasculitis itself is not contagious. An infection that triggers it, such as a hepatitis virus, may be transmissible. The route of transmission and precautions depend on the infection.

Can necrotizing vasculitis be cured?

Sometimes. A case caused by a medicine or treatable infection may resolve when the cause is removed or treated. Immune-mediated forms are more often controlled than permanently cured and may relapse after remission.

Does a positive ANCA test mean I have necrotizing vasculitis?

No. ANCA antibodies can support the diagnosis of certain vasculitis types, but they can occur in other situations and may be absent in someone who has vasculitis. Symptoms, urine results, imaging, and often a biopsy are also considered.

How long does treatment last?

Treatment length depends on the cause, severity, organs involved, response, and relapse risk. Initial treatment may be followed by maintenance therapy for an extended period. Monitoring often continues after medicines are reduced or stopped.

Can damaged organs recover?

Inflamed organs may improve when blood vessel inflammation is controlled. Recovery can take weeks or months. Severe loss of blood flow may leave permanent kidney, nerve, lung, skin, or other damage.

Sources

- Jennette JC, Falk RJ, Bacon PA, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides . Arthritis & Rheumatism . 2013;65(1):1-11.
- Chung SA, Langford CA, Maz M, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of ANCA-Associated Vasculitis . Arthritis & Rheumatology . 2021;73(8):1366-1383.
- Maz M, Chung SA, Abril A, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Polyarteritis Nodosa . Arthritis & Rheumatology . 2021;73(8):1384-1393.
- Hellmich B, Sanchez-Alamo B, Schirmer JH, et al. EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update . Annals of the Rheumatic Diseases . 2024;83(1):30-47.
- Kidney Disease: Improving Global Outcomes. KDIGO 2024 Clinical Practice Guideline for the Management of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis . Kidney International . 2024;105(3S):S71-S116.
- National Institute of Arthritis and Musculoskeletal and Skin Diseases. Vasculitis: Diagnosis, Treatment, and Steps to Take . National Institutes of Health. Reviewed November 2022.