

## RHEUMATOID VASCULITIS

### What is rheumatoid vasculitis (RV)?

Rheumatoid vasculitis (RV) is a rare but serious complication of rheumatoid arthritis (RA)—an inflammatory disease that causes severe joint pain and damage, and can move beyond the joints to other parts of the body. When RA affects the blood vessels, it is called rheumatoid vasculitis. RV causes inflammation and narrowing of the small- and medium-sized blood vessels, and most commonly affects the skin, nerves, fingers, and toes. However, it can involve virtually any organ in the body. People with RA who smoke may have a higher risk of developing this complication.

The primary symptoms of RV include skin rash; small pits on the fingertips or sores and redness around the nail; and numbness, tingling, and pain of the hands and feet. Because RV is systemic (affecting the whole body), it can also affect the heart, kidneys, gastrointestinal tract (GI), brain or lungs. RV resembles other forms of vasculitis, including polyarteritis nodosa and cryoglobulinemia.

With early diagnosis and treatment, it is possible to control symptoms of RV and prevent or minimize organ damage. Treatment depends on disease severity and involvement of other organs. Mild disease affecting the skin and nerves is typically treated with corticosteroids such as prednisone, and/or medications that suppress the immune system. More serious organ involvement may require higher doses of steroids, or other powerful agents that control inflammation.

Treatment decisions should take into account the underlying RA. Drugs taken for RA may improve both arthritis and vasculitis symptoms. Even with effective treatment, RV and RA are chronic diseases, so ongoing medical care is necessary.

### Causes

The cause of RV 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. Researchers believe a combination of factors may set the inflammatory process in motion, including genetics, drug reactions, or an infection or virus.

A number of factors point to the likelihood of a person with RA developing RV:

- Long-standing, severe RA, for 10 or more years
- High concentrations of “rheumatoid factor” antibodies, and the levels of certain other proteins in the blood
- Felty’s syndrome, a complication of RA characterized by an enlarged spleen and an abnormally low white cell count
- The presence of rheumatoid nodules, firm lumps under the skin around the joints of RA patients
- Smoking cigarettes, which may damage the lining of the blood vessels and predispose people to developing RV

### Who gets rheumatoid vasculitis?

RV is rare, occurring in less than 1 percent of those who have RA, while RA is considered common and affects millions of people annually. It is estimated that 400 people per million develop RA per year,

## RHEUMATOID VASCULITIS

compared to fewer than 4 people per million who develop RV. It appears that men are more likely to develop RV than women, and it can affect people from any ethnic background.

### Symptoms

RV mostly affects the skin, peripheral nerves, and the fingers and toes, but it can affect any organ and cause numerous symptoms. General symptoms of RV include fever, fatigue, loss of appetite and weight loss, which are common symptoms of RA as well. Other symptoms include:

- Small pits in the fingertips, sores/ulcers around the fingernails
- A painful red rash, especially on the legs, or purplish bruises
- Tissue death (necrosis) of the fingers and toes if blood flow becomes totally blocked
- Numbness, tingling and pain, especially in the fingers and toes
- Muscle weakness
- Chest pain
- Abdominal pain

### Complications

Although it is uncommon, serious, even life-threatening, complications can occur if RV affects major organs such as the heart, kidneys, lungs, GI tract or brain. These include:

- Heart attack
- Stroke
- Kidney failure
- Restriction of blood supply to the bowels
- Atherosclerosis

### Diagnosis

There is no single test to diagnose RV. Your doctor will consider a number of factors, including a detailed medical history; physical examination; laboratory tests (blood/urine); and specialized imaging studies. A tissue biopsy is typically required to make a definitive diagnosis.

Your doctor may suspect RV based on the characteristic symptoms of rash, numbness and tingling of the extremities, skin ulcers, chest pain or other symptoms. Diagnosis also involves ruling out diseases that cause similar symptoms, including other forms of vasculitis (polyarteritis nodosa and cryoglobulinemia); diabetes; and atherosclerosis, which can also cause narrowing of blood vessels.

Depending on organs affected, the following diagnostic tests, or others, may be ordered:

- **Blood tests:** To detect abnormal levels of antibodies and proteins in the blood, which can indicate inflammation. People with RV typically have elevated levels of rheumatoid factor antibodies, C-reactive protein (CRP), and erythrocyte sedimentation rate (ESR, or “sed rate”).
- **Imaging:** Chest X-ray; computed tomography (CT) or magnetic resonance imaging (MRI) scans of brain, chest or abdomen; angiogram (X-ray of blood vessel after injection of contrast dye)
- **Electrocardiogram (ECG):** To check heart rhythm

## RHEUMATOID VASCULITIS

- **Endoscopy:** An examination of the esophagus, stomach and portions of the intestine with a lighted, flexible instrument called an endoscope
- **Nerve conduction (EMG):** To test nerve function
- **Tissue biopsy** (usually required to confirm the diagnosis): This surgical procedure removes a small tissue sample from an affected vessel (skin ulcer, nerve or muscle), which is examined under a microscope for signs of inflammation or tissue damage.

### Treatment

The course of treatment for RV depends on the severity of vasculitis and organs involved. Any treatment decisions should take into account the underlying RA and how it is being treated. Drugs taken for RA may improve both joint symptoms and the vasculitis.

For mild symptoms of RV, such as fingertip sores, treatment involves protecting the area to prevent infection and use of corticosteroids, such as prednisone. For more widespread disease, corticosteroids may be used in combination with immunosuppressants, such as methotrexate or azathioprine. More serious organ involvement may require higher doses of steroids, cyclophosphamide (a chemotherapy-type drug), or biologic agents such as rituximab, abatacept, or others. Biologics are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation.

Research has shown that RV is associated with smoking. If you have RA and are a smoker, stopping smoking may reduce the risk of developing RV.

### Side effects

The medications used to treat RV and RA have potentially serious side effects, such as lowering the 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, relapses can occur with RV. If your symptoms return, or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

### Your medical team

Effective treatment of RV 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); neurologist (nerves); ophthalmologist (eyes); gastroenterologist (digestive system); (cardiologist (heart); pulmonologist (lungs); 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 your

## **RHEUMATOID VASCULITIS**

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 if necessary 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, be sure to speak with your medical team. 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 rheumatoid vasculitis**

Living with a serious health condition such as RV 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**

Rheumatoid vasculitis is one of the most serious complications of long-standing RA, and prognosis depends on disease severity and organ involvement. Some patients have milder symptoms while others can develop serious complications including heart failure, kidney failure and stroke, among others. Early diagnosis and treatment are essential in controlling the disease and preventing damage to blood vessels, vital tissues and organs. Not smoking may reduce the risk of developing RV.

The incidence of RV has decreased over the past 40 years due to improved RA treatments, including widespread use of biologic therapies.

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](http://www.vasculitisfoundation.org/research)

## **RHEUMATOID VASCULITIS**

### **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](http://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](https://www.facebook.com/groups/vasculitisfoundation)

## **RHEUMATOID VASCULITIS**

### **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](http://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](http://www.rarediseasesnetwork.org/cms/vcrc)

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