

What is Rheumatoid Vasculitis?

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.

RV is a form of vasculitis, which is an inflammation of the blood vessels.

RV most commonly affects the small vessels of the skin and peripheral nerves.

Causes

The cause of RV 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.

It is believed that a combination of factors may set the inflammatory process in motion, including genetics, drug reactions, or an infection.

There are a number of factors that 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, which is 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
- Erosions (damage) of the joints
- Smoking cigarettes, which may damage the lining of the blood vessels and predispose people to developing RV

Who Gets RV?

RV is rare and occurs in less than 1 percent of those who have RA. 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 and peripheral nerves, causing lesions more commonly seen in the fingers and toes.

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
- Leg ulcers
- 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 eyes and gastrointestinal tract. These include:

- Visual loss
- Restriction of blood supply to the bowels

Diagnosis

There is no single test to diagnose RV but diseases that cause similar symptoms will be ruled out. Your doctor will consider a number of factors, including:

- Detailed medical history
- Physical examination
- Laboratory tests (blood/urine)
- Specialized imaging studies
- A tissue biopsy to make a definitive diagnosis

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, and erythrocyte sedimentation rate (“sed rate”).

- **Imaging:** computed tomography or magnetic resonance imaging scans of chest or abdomen; angiogram (x-ray of blood vessel after injection of contrast dye) may be performed.
- **Electrocardiogram:** Used to check heart rhythm.
- **Endoscopy:** This is an examination of the esophagus, stomach and portions of the intestine with a lighted, flexible instrument called an endoscope.
- **Nerve conduction and needle test (EMG):** Used 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.

Your doctor may suspect RV based on the characteristic symptoms of rash, numbness and tingling of the extremities, skin ulcers, other symptoms, including other forms of vasculitis (polyarteritis nodosa and cryoglobulinemia); diabetes; and atherosclerosis, which can also cause narrowing of blood vessels.

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
- 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)
- Biologic agents such as rituximab, abatacept, or others. Biologics are:
 - Complex proteins derived from living organisms
 - Target certain parts of the immune system to control inflammation

RV has been shown to be 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
- Potential bone loss (osteoporosis), among others

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-stomach and bowels)
- 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 can be helpful to track medications, symptoms, test results and notes from doctor appointments.

Make a list of questions before your doctor's visit.

Bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it is 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 RV

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. 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

RV 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 severe neuropathy, 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.

(Revised October 2020)