Rheumatoid vasculitis (RV) refers to patients with rheumatoid arthritis, a chronic disease with painful inflammation of the joints, who also develop inflammatory disease in small and medium-sized blood vessels. RV most commonly occurs in the skin as venulitis or capillaritis, meaning the very smallest blood vessels are affected by inflammation from the disease. RV occurs in approximately 2 to 5 % of patients who have rheumatoid arthritis.

The **reason** why RV develops in some patients with rheumatoid arthritis and not others is not clear. Genetic factors may be involved. Viral infections and drug reactions have been suggested as causes of RV, but there is little research to support this. Some research suggests that long time use of drugs such as corticosteroids, gold compounds, penicillamine and azathioprine that are used to treat rheumatoid arthritis can cause the development of RV. However, this may not be true and difficult to determine because more use of these drugs is probably because of more severe or long standing rheumatoid arthritis, both of which may also be associated with the development of RV.

RV typically occurs in patients who have had rheumatoid arthritis for a long time. In one study, for example, the average time between the diagnosis of rheumatoid arthritis and the onset of RV symptoms was 13.6 years. Patients with rheumatoid arthritis seem more likely to develop RV when they have high rheumatoid factor levels (a specific laboratory finding for rheumatoid arthritis) and disease of at least one year's duration. Males with rheumatoid arthritis are more likely (2 to 4 times more likely) than females with rheumatoid arthritis to develop RV.

The manifestations of RV can involve many of the body's organs, including the skin, nerves to the hands and feet, blood vessels of the fingers and toes, and the eyes. Skin vasculitis is the most common manifestation of RV, occurring in as many as 90% of patients. Inflammation of the small blood vessels in the skin results in the development of red spots on the skin. When the eyes are involved, there is usually inflammation of the white part of the eye (scleritis).

The heart can also be affected by the disease, which can cause inflammation of the external part of the heart (pericarditis) and abnormal heart rate (arrhythmia). These symptoms put these patients at a higher risk for having a heart attack (myocardial infarction).

Patients with rheumatoid arthritis should see a physician if they develop new or worsened symptoms such as weight loss, fever, and lack of energy, any new symptoms beyond the usual joint symptoms. A blood test for specific antibodies that are directed against the inner layer of blood vessels (endothelial cells) are present in approximately 75% of patients with RV compared to only 15to 20% of those with rheumatoid arthritis alone. Therefore, this blood test may be checked regularly in patients with any of these new or worsened symptoms.

Diagnosis: Many of the drugs used to treat RV have a number of side effects; therefore it is important to be sure of the diagnosis before treatment is started (see treatment section below). The diagnosis of RV almost always requires a biopsy of tissue affected by the disease; an inflamed nerve or a kidney if there are clinical signs of kidney involvement, for example.

In rare cases RV may affect large blood vessels. If this happens or your doctor thinks it may have happened, then ‘pictures’ will be taken so that the vessels can be evaluated**.** Some of these pictures require that you drink something called ‘contrast’ material. This material shows up on the picture and helps to show different parts of the inside of your body. This test is called

contrast angiography and is especially useful to help determine the location and appearance of large vessels that may be affected by the disease.

Biopsy-proven RV, even if only in one organ, requires aggressive therapy. The limited data specifically related to RV suggest that most such patients should be treated with the same or similar drugs that are used in other primary systemic vasculitides such as combination therapy with cytotoxic drugs (usually cyclophosphamide) and corticosteroids. Cyclophosphamide given through the veins (intravenous) once a month has been used with success, although daily oral therapy with the same drug may also be effective.

A variety of different types of corticosteroid **treatment** have been used. Patients with aggressive RV disease are usually begun on pulse methylprednisolone (corticosteroids given through the veins once a day for several days) followed by daily oral prednisone. Other drugs have been explored in patients with RV. Some patients have done well with azathioprine and corticosteroids. However azathioprine may be better used to maintain a remission after initial cyclophosphamide therapy helps to control the disease and its symptoms. Methotrexate and tumor necrosis factor (TNF), also known as infliximab, have also been used. However, some patients have developed RV while on these drugs for the treatment of rheumatoid arthritis. Since RV most often occurs when there is very active rheumatoid arthritis, aggressive treatment usually helps to control symptoms of both rheumatoid arthritis and vasculitis.

Supportive care is also very important. Smoking has been associated with an increased risk for rheumatoid arthritis and for RV. Therefore smoking cessation is essential in any rheumatoid arthritis patient, especially those with RV. Good skin care may also prevent infectious complications of skin rashes in RV.

Limited data are available concerning the outcome of patients with RV, although they usually have worse and more ongoing symptoms than those with rheumatoid arthritis who do not have RV.