[Raajit K. Rampal](#)

Director, Center for Hematologic Malignancies; Director, Myeloproliferative Neoplasms Program

Ellin
Berman

Attending Physician

Michael J.
Mauro

Attending Physician

‹ ›

New Patient Appointments

Call 646-497-9154
Available Monday through Friday, 8:00 a.m. - 6:00 p.m. (Eastern time)

Make an Appointment

Back to top ^

Polycythemia Vera

Polycythemia vera develops slowly when bone marrow produces too many red blood cells. This can lead to a thickening of the blood. The condition is thought to be caused in part by a mutation in a gene called *JAK2. Normally, JAK2* regulates the production of different types of blood cells, keeping them in balance. More than 95 percent of people with polycythemia vera have a mutation in *JAK2* that leads to the production of too many red blood cells.

Scientists think people develop the mutation, rather than inherit it from a parent. Our research, however, suggests that people can inherit a genetic abnormality that increases their risk of developing a *JAK2* mutation.

Polycythemia vera usually doesn't cause any signs or symptoms in its early stages. After progressing, it may cause headaches, shortness of breath, bleeding, dizziness, itchiness, or an enlarged spleen. The disease can also increase the likelihood of developing blood clots and the risk of stroke.

Polycythemia vera is usually diagnosed through a blood test, if the test reveals abnormal levels of red blood cells. To confirm the diagnosis, we also may recommend a bone marrow biopsy and a test for the specific genetic mutation in *JAK2*.

Polycythemia Vera Treatment

Polycythemia vera cannot be cured. Treatment is given to manage the disorder as a chronic condition. The standard treatment is phlebotomy (the removal of a specific amount of blood) on a regular basis, sometimes combined with other therapies.

For some people, such as those who have had blood clots, chemotherapy is used instead of phlebotomy to stop the excess production of red blood cells. This may include the drug hydroxyurea (Hydrea®). It limits the bone marrow's ability to make blood cells.

Interferon may be used to stimulate the immune system and slow the production of red blood cells. For itchy skin, our doctors may prescribe medication, such as an antihistamine.

People who have taken hydroxyurea but have had either a poor response or side effects can be treated with ruxolitinib (Jakafi®), a drug that targets the *JAK2* mutation.

Clinical trials for people with polycythemia vera using new drugs are currently ongoing.

Some people with polycythemia vera develop a blood disorder called myelofibrosis. Others develop acute myeloid leukemia. We are researching new treatment options for people with myelofibrosis or acute myeloid leukemia that developed after polycythemia vera who are not responding to standard therapies.

Back to top ^

Myelofibrosis

Myelofibrosis happens when bone marrow cells called fibroblasts make too much fibrous (scar) tissue within the bone marrow. As a result, fewer blood-producing cells are created and they can be destroyed more rapidly. This causes anemia (low iron), a low platelet count, and a tendency to develop infections.

Myelofibrosis is most common in people between the ages of 50 and 70. It often produces few symptoms at first. When anemia develops, it can cause fatigue, weakness, and abdominal pain from an enlarged spleen. Myelofibrosis can occur by itself or with other blood disorders.

Myelofibrosis Treatment

There are a variety of treatments for myelofibrosis. Some people do not require any active treatment and may be followed closely. This is called active surveillance. Other people may require treatment. The decision depends on the symptoms and how the disease acts. Ruxolitinib (Jafaki®) is a drug that targets *JAK2* and other associated mutations. It can reduce the size of the spleen and lessen many myelofibrosis symptoms.

People with myelofibrosis often have anemia. This can be treated with blood transfusions. There are a variety of ways to treat anemia as well. These include the drugs danazol, thalidomide (Thalomid®), and lenalidomide (Revlimid®), as well as the hormone erythropoietin.

Other drugs that treat myelofibrosis include interferon and hydroxyurea (Hydrea®).

The only potential cure for myelofibrosis is a bone marrow transplant. Not everyone with the disease is able to have this procedure.

Some people with myelofibrosis develop acute myeloid leukemia. At MSK, we are committed to improving outcomes for people with myelofibrosis and people with acute myeloid leukemia that has developed after myelofibrosis. We are exploring new treatment options for people with these conditions and are working to translate new ideas from our research laboratories into innovative clinical trials.

Back to top ^

Essential Thrombocythemia

Essential thrombocythemia results when the bone marrow produces too many platelets. The condition can cause blood clots throughout the body that can affect the vital organs.

To make a diagnosis of essential thrombocythemia, our doctors usually do a bone marrow biopsy. In this test, a needle is inserted into the bone to draw a sample of marrow for analysis under a microscope.

Chemotherapy can be used to stop the excess production of platelets. This may include the drugs hydroxyurea (Hydrea®), interferon, or anagrelide (Agrylin®), which change the bone marrow's ability to make blood cells.

Some people with essential thrombocythemia develop myelofibrosis . We are exploring new treatment options for essential thrombocythemia and for people who develop myelofibrosis after essential thrombocythemia and are not responding to standard therapies.

Back to top

Eosinophilia

People with eosinophilia have an abnormally high number of eosinophils, a type of white blood cell, in their blood. High levels of eosinophils show that the body may be reacting to an allergen, a parasite, or substances produced by some cancers, such as Hodgkin's disease and chronic myeloid leukemia.

Symptoms of eosinophilia, if they occur, are usually symptoms of the underlying allergy, parasite, or cancer. At MSK, our doctors often treat the illness by addressing the root cause.

Back to top

Chronic Neutrophilic Leukemia

Chronic neutrophilic leukemia is a rare myeloproliferative disorder. It occurs when too many neutrophils, a type of white blood cell, are made in the bone marrow. These cells spill out into the circulating blood and accumulate in the liver and spleen, which can become enlarged as a result.

New genetic insights into this disease have led to new treatment possibilities. Treatment options may include chemotherapy drugs, such as hydroxyurea (Hydrea®), or new targeted drugs, such as ruxolitinib (Jafaki®).

Back to top

PREVIOUS
Treatment for Hairy Cell Leukemia

NEXT
Living Beyond Leukemia