

Polycythemia Vera (PV)

The red color of blood is attributed to the presence of hemoglobin, which is contained within red blood cells. Hemoglobin carries oxygen from the lungs to various parts of the body. Approximately 15 milligrams of hemoglobin can be found in each milliliter of blood. Red blood cells are produced in the bone marrow, the soft tissue inside bones. When individuals suffer from conditions like lung problems, smoking addiction, or heart defects, leading to inadequate oxygen supply to different body parts, the kidneys secrete a hormone called Erythropoietin in large quantities. Erythropoietin acts on bone marrow stem cells to stimulate the production of more red blood cells, a process known as secondary polycythemia. In contrast, individuals with polycythemia vera experience excessive production of red blood cells by the bone marrow without the need for Erythropoietin stimulation, making it a type of cancer. However, it is not life-threatening like other aggressive cancers, and it does not necessitate powerful chemotherapy.

Polycythemia vera is usually caused by a mutation in the JAK-2 gene, although the exact cause of this mutation remains unknown.

With an increase in hemoglobin levels, the blood's density rises, transforming the blood from a fluid resembling buttermilk to a thicker consistency like thin curd. Consequently, there is an increased risk of blood clots

forming in the blood vessels. As the blood flow becomes sluggish, blood vessels may become congested, leading to swelling, rupture, and bleeding.

Reduced blood circulation can result in symptoms such as headaches, lack of concentration, mental fatigue, dizziness, and visual disturbances, including blackouts. Redness of the face and eyes may also be observed in patients. Additionally, some individuals may experience an enlarged spleen, itching in skin, and a burning sensation in the fingers. In cases where blood clots form in the veins, the legs get swollen. In severe situations, cardiac arrest resulting from block in coronary vessels or stroke can occur.

Approximately 20% of patients may develop bone marrow failure after around 10 years, which is known as the Spent Phase. In a small number of people, polycythemia vera can transform into a more serious form of blood cancer known as AML (acute myeloid leukemia).

To confirm the presence of PV disease, a doctor will conduct several tests. The initial step involves measuring the amount of Erythropoietin hormone. Subsequently, a bone marrow test and a JAK-2 test are performed.

PV patients are categorized as high-risk or low-risk based on factors such as the patient's age, presence of blood clots in

veins, and the amount of blood that needs to be drained. Low-risk patients are prescribed aspirin to prevent blood clots. To maintain blood PCV/HCT levels below 45% in men and 42% in women, frequent blood removal is required. This process is similar to blood donation and is conducted in the blood bank. It is crucial to follow heart-healthy practices such as brisk walking, controlling blood pressure, cholesterol, and sugar levels, managing obesity, and quitting smoking/alcohol consumption. High-risk patients may be prescribed Hydroxyurea tablets along with the aforementioned measures. Regular blood tests are conducted every two to three months to monitor the effectiveness of Hydroxyurea. The drug dosage is adjusted based on its impact. In cases where Hydroxyurea is not suitable, an injection called Interferon may be administered. If blood clots persist or other disease-related symptoms persist despite medication, Ruxolitinib may be prescribed. In some cases, PV can progress to other types of cancer, such as AML and Myelofibrosis, which require different treatments.

Many people with PV disease may not be aware of their condition until they experience blood clots in their veins. In such cases, treatment for PV disease must be combined with treatment for the clot.

When detected early and treated appropriately, patients with PV can live an average of more than ten years.

Dr. Girish Kamat MD, DNB (Hematology)

Professor,
Department of Hematology,
SDM College of Medical Sciences and Hospital,
Sri Dharmasthala Manjunatheshwara University,
Dharwad- 580008

Patients often worry about complications arising from repeated blood draws. One should note that, the side effects of repeated blood draws are minimal compared to the high risk associated with increased hemoglobin level. Patients need not be overly concerned about this.

The side effects of hydroxyurea tablets are closely monitored by the doctor every two to three months, and appropriate measures are taken if adverse effects are observed. Aspirin tablets typically have no significant side effects, but they may need to be discontinued for four to five days before undergoing surgery.

Patients with PV should reduce their intake of iron-rich foods such as meat, greens like spinach, fish, nuts, etc., as well as foods with high fat content.

PV disease is neither hereditary nor transmitted from person to person. Ongoing research is focusing on the development of new drugs for the treatment of this disease, and it is likely that new treatment options will become available in the coming years.

For additional information about PV disease beyond what is provided here, patients can consult their treating doctor to obtain accurate and comprehensive details.

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