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MYELOPROLIFERATIVE NEOPLASMS

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ABSTRACT

Introduction- The abnormal proliferation of one or more terminal myeloid cell lines in the peripheral blood gives rise to a heterogeneous group of disorders called myeloproliferative neoplasms. Definition - Myeloproliferative neoplasms (myeloproliferative disorders) are rare blood cancers that occur when your body makes too many red blood cells, white blood cells or platelets. Types of myeloproliferative neoplasms- Chronic eosinophilic leukemia, Chronic myelogenous leukemia, Chronic neutrophilic leukemia, Myeloproliferative neoplasm, unclassifiable. Symptoms- Splenomegaly feels like fullness, pressure or discomfort below your ribs on your left side, where your spleen is located. Causes- Mutations associated with Janus kinases, Mutations associated with the *MPL* gene or *CALR* gene, Chromosome errors. Diagnosis- CBC, PBS, bone marrow biopsy, genetic test. Management and treatment - Chronic eosinophilic leukemia, Chronic myelogenous leukemia, Chronic neutrophilic leukemia, Essential thrombocythemia, Polycythemia vera, Primary myelofibrosis. Conclusion- With careful monitoring and treatment, many people live for several years. There's no single prognosis or expected outcome for these conditions

KEY WORDS – Introduction, definition, types, causes, diagnosis, management and treatment, conclusion

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INTRODUCTION

The hematopoietic process is determined by the bone marrow environment, growth factors, and transcription factors. The abnormal proliferation of one or more terminal myeloid cell lines in the peripheral blood gives rise to a heterogeneous group of disorders called myeloproliferative neoplasms.

DEFINITION

Myeloproliferative neoplasms (myeloproliferative disorders) are rare blood cancers that occur when your body makes too many red blood cells, white blood cells or platelets. Your healthcare provider can work with you to relieve symptoms, slow disease progression and prevent complications.

TYPES OF MYELOPROLIFERATIVE NEOPLASMS

Depending on the type of myeloproliferative neoplasm, your bone marrow may make too many red blood cells, white blood cells, platelets or a combination of cell types. The cells often behave differently from healthy blood cells.

OTHER TYPES OF MYELOPROLIFERATIVE NEOPLASMS

- **Chronic eosinophilic leukemia (CEL)** involves the overproduction of white blood cells called eosinophils. Usually, it progresses slowly. In rare cases, CEL can become a serious form of cancer called acute myeloid leukemia (AML). CEL is also called hypereosinophilic syndrome.
- **Chronic myelogenous leukemia (CML)** involves an overproduction of white blood cells called granulocytes. These cells accumulate, making it harder for your bone marrow to make other blood cells your body needs.
- **Chronic neutrophilic leukemia (CNL)** involves an overproduction of white blood cells called neutrophils.
- **Myeloproliferative neoplasm, unclassifiable (MPN-U)**, is a type of myeloproliferative neoplasm that doesn't fit into the other categories. It may involve an overproduction of various blood cell types, including white blood cells, red blood cells or platelets.

MYELOPROLIFERATIVE NEOPLASM SYMPTOMS

As your condition progresses, you may notice signs of an enlarged spleen (splenomegaly). Splenomegaly feels like fullness, pressure or discomfort below your ribs on your left side, where your spleen is located. While splenomegaly is a common symptom of most myeloproliferative neoplasms, it's a less common symptom of essential thrombocytopenia.

CHRONIC MYELOGENOUS LEUKEMIA AND CHRONIC NEUTROPHILIC LEUKEMIA

- Bone pain.
- Night sweats.
- Fever and fatigue.
- Bruising easily.
- Loss of appetite and weight loss.

ESSENTIAL THROMBOCYTHEMIA

- Bruising easily.
- Unexplained bruising or bleeding from your nose, mouth and gums.
- Bleeding from your stomach or intestines.
- Blood in your pee.

POLYCYTHEMIA VERA

- Headaches.
- Dizziness.
- Fatigue.
- Blurred or double vision.

PRIMARY MYELOFIBROSIS

symptoms of anemia (fatigue, weakness, shortness of breath)

. Pale skin.

- Night sweats.
- Fevers.
- Itchy skin.
- Abdominal fullness or filling up right away when you eat (early satiety).
- Weight loss.
- Bone pain.

CAUSES MYELOPROLIFERATIVE NEOPLASMS

- **Mutations associated with Janus kinases (JAK)**: Polycythemia vera, primary myelofibrosis and essential thrombocythemia often involve genetic mutations associated with a protein called Janus kinase 2 (JAK2). The mutation may cause cells to multiply out of control.
- **Mutations associated with the *MPL* gene or *CALR* gene**: People with essential thrombocythemia and primary myelofibrosis often have mutations in their *MPL* gene or *CALR* gene.
- **Chromosome errors**: People with chronic myelogenous leukemia (CML) have a specific error involving their chromosomes. A chromosome is a structure that contains genes. With CML, a piece of one chromosome swaps places with another chromosome, forming the "Philadelphia chromosome."

MYELOPROLIFERATIVE NEOPLASMS DIAGNOSIS

- **Complete blood count (CBC)**: This test measures all blood cell levels. In essential thrombocythemia, providers evaluate platelet levels. In polycythemia vera, they look for increased hemoglobin — the protein in red blood cells — as well as white blood cells and platelets.
- **Peripheral blood smear (PBS)**: This test can show abnormal cell shapes that may indicate a condition. Blood chemistry tests can detail how much of a specific type of chemical is in your blood (proteins, enzymes, glucose, etc.). These numbers provide clues about how your organs are functioning, which may suggest a myeloproliferative neoplasm.
- **Bone marrow biopsy**: Your provider may do bone marrow aspiration or bone marrow biopsy. In this test, they remove a sample of bone marrow to check your blood cells. Then, medical pathologists examine blood cells and tissue under a microscope, looking for differences between normal and abnormal cells. They'll see if you have an unusual number of stem cells. They'll also look for changes in chromosomes and other signs of genetic mutations that may indicate you have a specific type of myeloproliferative neoplasm.
- **Genetic testing**: Providers may analyze your blood cells, looking for changes in the genes that may affect blood cell production.

MANAGEMENT AND TREATMENT

- **Chronic eosinophilic leukemia**: Your provider will work to reduce your eosinophil levels with chemotherapy, corticosteroids or immunotherapy.
- **Chronic myelogenous leukemia**: The most common treatment is targeted therapy that prevents cells from multiplying out of control. Other treatments include chemotherapy, immunotherapy, radiation therapy and stem cell transplants.
- **Chronic neutrophilic leukemia**: Treatments may include chemotherapy, immunotherapy and stem cell transplants.
- **Essential thrombocythemia**: If you don't have symptoms, your provider may choose to monitor your condition closely instead of prescribing treatments. If you have symptoms, you may need to take a

treatment that prevents cells from multiplying out of control. You may need to take medicine to reduce your risk of blood clots or to prevent your bone marrow from making too many platelets.

- **Polycythemia vera**: Phlebotomy is the most common procedure to treat polycythemia vera. Your healthcare provider will regularly remove blood (like a blood draw) to reduce your blood volume and remove excess red blood cells. If you have symptoms, you may have targeted therapy that prevents cells from multiplying out of control. You may also take medicine to reduce your risk of blood clots (like aspirin) or to reduce your number of red blood cells.
- **Primary myelofibrosis**: Your provider may decide to monitor your condition closely if you aren't experiencing symptoms. Treatments may include procedures or drugs to treat anemia. For example, you may need a blood transfusion if your bone marrow isn't making enough red blood cells. You may take medicines that stimulate your bone marrow to produce more blood cells. Other treatments may include targeted therapy, chemotherapy, immunotherapy, radiation therapy and stem cell transplant.

CONCLUSION

With careful monitoring and treatment, many people live for several years. There's no single prognosis or expected outcome for these conditions. In general, people diagnosed with MPN are alive five years later.

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