Chronic Myeloproliferative Disorders/Myeloproliferative Neoplasms

Chronic Myeloproliferative Disorders, also called Myeloproliferative Neoplasms, are a group of diseases in which the bone marrow makes too many blood cells. These can be red blood cells, white blood cells, or platelets. The bone marrow is the soft spongy center of bone. It is a factory for making new blood cells from immature blood cells, called stem cells. In myeloproliferative disorders, too many stem cells make one or more types of blood cells. The disease gets worse slowly as the number of extra blood cells build up in the blood and bone marrow. This may cause bleeding problems, anemia, infections, fatigue or other symptoms.

What are the types of Chronic Myeloproliferative Disorders?

There are four types of Chronic Myeloproliferative Disorders. The type is based on the kind of extra blood cells made by the bone marrow. This usually leads to too many red blood cells, white blood cells, or platelets in the bone marrow and blood stream. They include:

- Chronic myelogenous leukemia– a cancer of the blood cells in which there are a large number of white blood cells made by the bone marrow that are not normal and do not work correctly.
- Polycythemia vera: this is a condition where there are too many red blood cells in the blood. About 95% of these patients have a change in a cell protein called JAK2 which controls how blood is made.
- Primary myelofibrosis: this condition causes scar tissue to grow inside the bone marrow. It may cause your bone marrow to make too few numbers of all blood cells. About 50% of patients with this condition have a change in a cell protein called JAK2. JAK2 controls how blood is made.
- Essential thrombocythemia: this is a condition where there are too many platelets in the blood. The platelets are “sticky” and do not always work well. Abnormal platelets can lead to bleeding or clotting problems. About 50% of these patients have a change in a cell protein called JAK2 which controls how blood is made.

Sometimes chronic myeloproliferative disorders become Acute Leukemia, a condition where too many abnormal white blood cells are made and do not work properly. Acute Leukemia grows quickly without treatment.

What are the symptoms of myeloproliferative disorders?

The symptoms depend on the type of disorder you have. In general, you may see some of the following symptoms:

- bleeding problems
• repeated headaches
• anemia
• shortness of breath
• chest pain
• infections
• fatigue
• enlarged spleen, an organ in the upper left of the abdomen
• enlarged liver
• night sweats
• bone pain
• weight loss for no clear reason
• itching

Each of these symptoms alone can be caused by other medical conditions. Call your treatment team if you have any of these symptoms.

**How are Chronic Myeloproliferative Disorders diagnosed?**

- Full medical history by your doctor
- Full physical exam with attention paid to enlarged liver and spleen
- **Blood tests:** Complete Blood Count (CBC) to check the number and kinds of white blood cells, red blood cells and platelets in your blood. Learn more about understanding your blood counts.
- **Blood Cell Examination:** a test on the blood cells where the blood cells are washed with a colored dye and looked at through a microscope, also called a blood smear. Blood is checked see whether there are abnormal shapes to the red blood cells, such as teardrop shape instead of round. They also look to see if there are early immature cells called blast cells.
- **Biopsy:** a test to remove a very small piece of tissue to look for cancer cells in your bone marrow. The tissue usually comes from the back of your hipbone. The test is done with numbing medicine to reduce the discomfort of the procedure. The tissue removed is looked at under the microscope by a pathologist (tissue doctor) to check for abnormal cells. A bone marrow aspirate and biopsy are usually done together in the office.

  **Bone marrow aspiration:** using a hollow needle to remove the liquid sample of bone marrow into a syringe

  **Bone marrow biopsy:** capturing the solid piece of the bone marrow that stays in the bone marrow needle after it is removed from the patient. This solid piece is placed in a jar.
• **Cytogenetic Analysis**: a test on the sample of bone marrow or blood to determine the make-up of the genetic material inside of the cells. These findings are important in the planning of treatment.

• **JAK2 gene mutation test**: lab test done on bone marrow or a blood sample to look for a change (mutation) in a piece of genetic material in your cells (a gene) call JAK2. This is often found in three of the Myeloproliferative Disorders: polycythemia vera, essential thrombocytopenia, or primary myelofibrosis.

If you have polycythemia vera, you may need to have additional blood tests such as an EPO test, which checks erythropoietin (a hormone) levels. You may also need testing for iron, folate, and vitamin B12 levels.

**What is the Treatment for Myeloproliferative Disorders?**

The treatment for myeloproliferative disorders depends on the type of disorder that you have and your clinical condition. There are many treatment options that range from:

• low dose chemotherapy (such as hydroxyurea)
• a pill that is targeted against JAK2 (called ruxolitinib)
• surgery to remove your spleen or
• radiation therapy to the spleen.
• Removal of a pint of blood periodically to reduce the high red blood cell levels (polycythemia vera)
• CML is treated with oral chemotherapy pills (please see CML section)

Some patients may be appropriate for a donor bone marrow or stem cell transplant. You may also be eligible for a clinical trial of a new treatment.

**Clinical Trials**

A Clinical Trial is a research study of new drugs, new combinations of drugs or already approved drugs being studied to treat patients in new/different ways. They may include new drug doses or new ways (schedules) to give the drugs. Clinical trials are run under rigid guidelines. Their purpose is to help find out whether new cancer treatments are safe and effective or better than the standard (current) treatment. At Massachusetts General Hospital, we have several clinical trials open for the treatment of leukemia, which use the latest in cancer treatments.
If you have any questions or would like to speak with one of our physicians, please call the Center for Leukemia at Massachusetts General Hospital at 617 724 1124. You can speak to a senior Leukemia physician 24 hours a day/ 7 days a week by calling Dr. Karen Ballen at 617-724-5700 beeper 31343