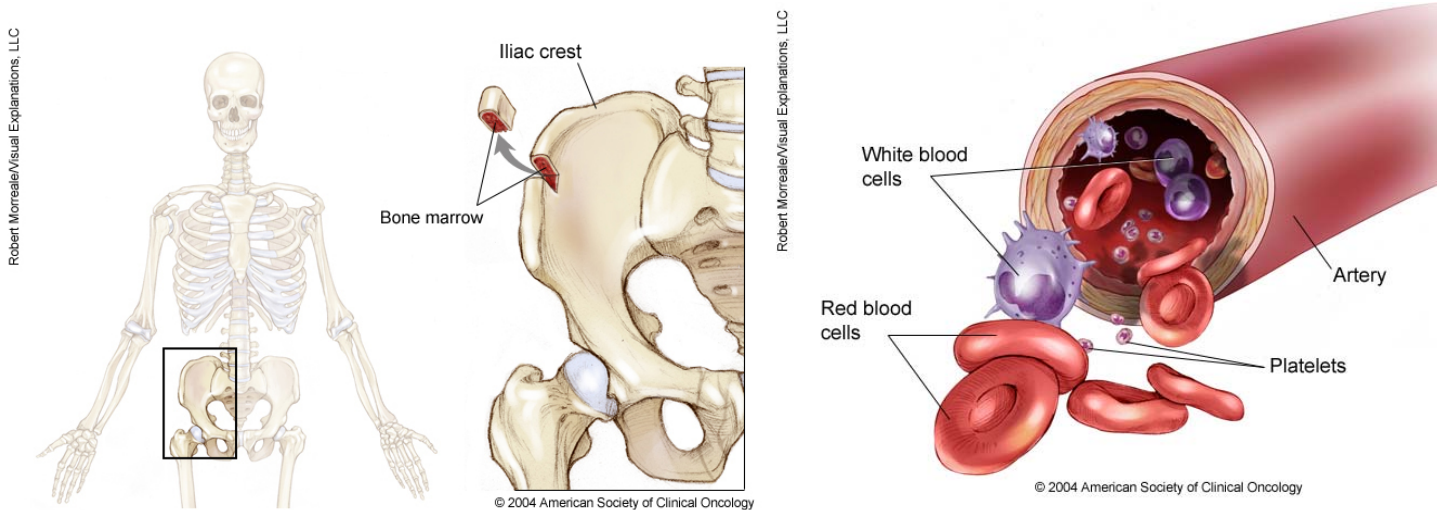


## CHRONIC MYELOGENOUS LEUKEMIA

Chronic myelogenous leukemia (CML) is a disease in which the bone marrow makes too many white blood cells. It is usually a slowly progressing blood and bone marrow disease that normally occurs during or after middle age, and rarely occurs in children. Normal bone marrow produces stem cells that become red blood cells, white blood cells or platelets. Red blood cells carry oxygen and nutrients to all tissues in the body. White blood cells fight infection and are an important part of the body's immune system. Platelets help stop bleeding by causing blood clots to form. CML affects the bone marrow stem cells so that they develop into a type of white blood cell called a granulocyte. Some of these bone marrow stem cells never become mature white blood cells. These are called blasts. The granulocytes and blasts eventually crowd out the red blood cells, white blood cells, and platelets in the bone marrow. This may result in infections, anemia, and easy bleeding, as well as bone pain and pain or a feeling of fullness below the ribs on the left side. The fullness feeling is due to the extra white blood cells collecting in the spleen, causing it to enlarge.



### **SYMPTOMS TO REPORT** include:

- \*Feeling very tired
- \*Weight loss for no known reason
- \*Night sweats
- \*Fever
- \*Pain or a feeling of fullness below the ribs on the left side

These symptoms may be caused by CML or other conditions. A health care provider should be consulted if any of the problems occur. Sometimes CML does not cause any symptoms at all.

Most people with CML have a gene mutation (change) called the Philadelphia chromosome. Every cell in the body contains genetic material called DNA that determines how the cell looks and acts. DNA is contained inside chromosomes. In CML, part of the DNA in the blood cells moves from one chromosome to another chromosome. This change is called the "Philadelphia chromosome." It results in the bone marrow making an enzyme, called bcr-abl tyrosine kinase, which causes too many stem cells to develop into white blood cells (granulocytes or blasts). The Philadelphia chromosome is not passed from parent to child.

## DIAGNOSING AND STAGING

There is no standard staging system for CML. Staging is the process used to find out how far the cancer has spread. CML is classified by phase. Treatment is planned once the phase is determined. The following tests and procedures may be used to diagnose CML and determine the phase of the disease.

**Physical exam** of entire body, including health habits and past illnesses and treatments

**Blood tests** of a complete blood count (CBC) to measure the counts of white and red blood cells; and blood chemistry studies to check levels of certain substances in the blood

**Bone marrow biopsy** is the removal of a small piece of bone, bone marrow, and blood through a needle inserted into the hipbone or breastbone. The cells will be examined by a pathologist for signs of cancer and changes.

**Cytogenetic or fluorescence in situ hybridization (FISH) analysis** is a test to examine blood or bone marrow cells under a microscope to look for certain changes in the chromosomes, such as the Philadelphia chromosome. This test is also used to measure how well the treatment for CML is working.

## PHASES

The number of blast cells in the blood and bone marrow and the severity of symptoms determine the phase of the disease.

**Chronic phase:** fewer than 10% of the cells in the blood and bone marrow are blast cells

**Accelerated phase:** 10% to 19% of the cells in the blood and bone marrow are blast cells

**Blastic phase:** 20% or more of the cells in the blood or bone marrow are blast cells. Blast crisis is caused by having very few normally functioning red and white blood cells and platelets. Symptoms are tiredness, fever, and an enlarged spleen.

**Relapsed Chronic Myelogenous Leukemia:** number of blast cells increases after symptoms have initially disappeared but then return.

## TREATMENT OPTIONS

Certain factors affect the chance of recovery and the choices for treatment. These include the patient's age, the phase of CML, the amount of blasts in the blood or bone marrow, the size of the spleen at the time CML is diagnosed, and the patient's general health. Six types of standard treatment are used.

**Tyrosine kinase inhibitor therapy** is used to block the enzyme that causes stem cells to develop into more white blood cells than the body needs. This is a medication that is taken by mouth. It has become the most common treatment for CML.

**Chemotherapy** uses drugs taken by mouth, injections through a vein or muscle, or placed directly into the spinal column or in an organ to stop the growth of cancer cells. Chemotherapy is usually only given to CML patients who have developed the blast phase of their disease.

**Biologic therapy** is a treatment that helps the patient's immune system to fight cancer. Substances made by the body or in a laboratory are used to boost, direct, or restore the body's natural defenses against cancer.

**High-dose chemotherapy with stem cell transplant** is a method of giving high doses of chemotherapy and replacing blood-forming cells that are abnormal or destroyed by the cancer treatment. Stem cells are immature blood cells that are removed from the blood or bone marrow of the patient or donor and then frozen or stored. The stem cells are thawed and given back to the patient through an infusion after the high-dose chemotherapy. The reinfused stem cells grow into and restore the body's blood cells.

**Donor lymphocyte infusion (DLI)** is a treatment that may be used after stem cell transplant.

Lymphocytes (a type of white blood cell) from the stem cell transplant donor are removed from the donor's blood and may be frozen or stored. The donor's lymphocytes are then given to the patient through one or more infusions. The donor's lymphocytes then attack the patient's CML cells.

**Surgery** to remove the spleen is also an option.