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Living with Chronic Myeloid Leukemia (CML)

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Chronic myeloid (or myelogenous) leukemia (CML) is one of four main types of leukemia that can affect adults. These 4 types of leukemia are classified as either *myeloid* or *lymphocytic* based on the type of cells affected, and either acute or chronic. The chronic forms of leukemia tend to take longer to develop than the acute forms of leukemia; and therefore, people with chronic leukemia can live with the disease for many years.

But there are some important things to know in order to manage life with this disease. Following a diagnosis of CML, it is important for you to understand your treatment options, the expected results of each option, possible side effects and what option may be best for you. An ongoing commitment to stick to your prescribed treatment plan and to discuss any worries or concerns that you might have with your healthcare team is key to managing CML. Also, it's necessary to work closely with your healthcare team who can help you understand the disease better, learn how the disease may progress and find out who you can reach out to for support.

What is CML? And Who's at Risk?

Accounting for almost 15% of all adult leukemia, CML is a rare form of leukemia that causes too many white blood cells to be formed and build up in the blood and bone marrow. These abnormal white blood cells also known as leukemia cells are formed in the bone marrow, which is the soft inner part of bones that contains immature blood-forming cells, or blood stem cells. If not treated, CML cells may later crowd out normal blood

cells causing disease progression.

CML can be diagnosed at any age, but mostly occurs in older adults, with almost half of the cases occurring in people 65 years and older. According to the American Cancer Society, the average age at diagnosis is 64 years, and the risk for CML increases with age. Additionally, CML seems to be more common in men than women. There are no other proven risk factors for CML.

How to Recognize CML

Because CML is generally a slow growing cancer, there may not be any telltale signs or symptoms during the early stages of the disease. In fact, many people have mild or no symptoms for months or years before detecting it. Often it is discovered during a physical or check-up—a routine blood test may show abnormal blood results, such as a high white blood cell count, or a low red blood cell and platelet count. Most people are diagnosed in this early stage (or *chronic* phase).

In the more progressive stage (or *accelerated* phase) of CML, there is an increase of immature white blood cells in the blood or bone marrow—this is the stage in which symptoms may be present. The most advanced stage of CML is the *blast* phase (also called acute phase or blast crisis). In blast crisis, symptoms are often present and chances for survival are significantly reduced.

The symptoms of CML can seem rather general and may include:

- Fatigue.
- Appetite loss.
- Weight loss.
- Fever.
- Pale skin.
- Extreme sweating during sleep.
- Pain or feeling full in the upper left side of the belly.

Doctors will initially perform a physical exam and order a blood test. He or she may then conduct bone marrow testing. Special tests are performed on the blood and/or bone marrow to look for the presence of a mutated gene.

What Causes CML?

CML is caused by a change, or mutation, in chromosomes. In fact, a piece of chromosome 22 breaks off and attaches to a section on chromosome 9, and vice-versa—a piece of chromosome 9 breaks off and attaches itself to chromosome 22. This results in a short chromosome 22 and long chromosome 9.

The mutated chromosome 22 is called the Philadelphia (Ph) chromosome (named for the city where it was discovered). This Ph chromosome then creates a new gene called *BCR-ABL*. The newly formed *BCR-ABL* gene causes cancer by leading to the production of a certain kind of protein (known as a *tyrosine kinase*) that is responsible for causing the immature blood-forming cells to increase in number and grow out of control. A small number people with CML do not have the Philadelphia chromosome in their blood, but they still have the cancer-causing gene *BRC-ABL*.

Treating CML

Breakthroughs in medicine research and better understanding of CML at the chromosomal level have led to the development of new therapies which target the protein (*tyrosine kinase*) responsible for the cells growing out of

control. The development of these targeted therapies (called *tyrosine kinase inhibitors*) has led to a significant increase in the survival rates for newly diagnosed CML patients over the past few years. These targeted therapies are now considered the standard treatment for CML; however, none of them cure CML, and so these drugs need to be taken indefinitely to continue to inhibit the protein made by the cancer-causing gene *BRC-ABL*. Additionally, if one therapy stops working, patients may need to switch therapies.

Besides targeted therapies, other treatments may also be used to treat CML. The use of other therapies depends on the stage of disease and other clinical factors. These treatments may include chemotherapy, radiation therapy, surgery, or stem cell transplant. Participating in a clinical trial for potential new CML treatments may also be an option for some people with CML. If you are diagnosed with CML, your healthcare provider will discuss your treatment options with you and help you decide on a treatment plan.

Managing Life with CML

Treatment for CML may remove or destroy the cancer in a few people. For most people, however, the cancer is not cured and continued treatment is needed. Whether your cancer has been cured or not, it is important to continually follow-up with your doctor so that you can be watched for signs of recurrence, progression, treatment resistance, or treatment side-effects.

If you are diagnosed or have CML, here are some other important points to think about:

- **Learn about CML.** Understanding the disease may encourage you to take your medications as prescribed and to take an active role in your own care.
- **Keep your medical appointments.** CML is usually a lifelong disease and requires regular follow-up and monitoring.
- **Ask questions.** Write down questions that you want to ask (e.g. what phase is my CML in, what are my treatment options, how long will treatment last, what are the potential side effects of treatment, how often will you test my blood or bone marrow).
- **Take medication as prescribed.** And do not stop taking your medication without consulting your doctor. Let your healthcare team know if you experience any changes in symptoms or any side effects* due to medication.
- **Be positive.** With close collaboration with your healthcare team and proper management, it may be possible for people with CML to live for many years.
- **Get support.** Sharing the stress of having cancer with others may help you feel that you are not alone. Online communities can help offer emotional support as well.

*If you think you're experiencing an adverse event with any medication, you should contact your healthcare provider right away and then report it to FDA MedWatch.

Keep in mind that your healthcare team can offer help with managing side effects and taking your medication as prescribed. They can tell you about other treatment options and clinical trials that may be available to you. Some providers may also be able to help you find places that offer support for patients who cannot afford to pay for expensive medications.

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