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Leukemia - B-cell Prolymphocytic Leukemia and Hairy Cell Leukemia - Treatment Options [1]

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ON THIS PAGE: You will learn about the different ways doctors use to treat people with HCL and PLL. To see other pages, use the menu on the side of your screen.

This section outlines treatments that are the standard of care (the best proven treatments available) for HCL and PLL. When making treatment plan decisions, patients are also encouraged to consider clinical trials as an option. A clinical trial is a research study to test a new approach to treatment to evaluate whether it is safe, effective, and possibly better than the standard treatment. Clinical trials may test such approaches as a new drug, a new combination of standard treatments, or new doses of current therapies. Your doctor can help you review all treatment options. For more information, see the [Clinical Trials](#) [3] and [Latest Research](#) [4] sections.

Treatment overview

In cancer care, different types of doctors often work together to create a patient's overall treatment plan that combines different types of treatments. This is called a [multidisciplinary team](#) [5].

The goal of treatment for HCL and PLL is remission. Remission is the absence of leukemia symptoms and is also called "no evidence of disease" or NED. Descriptions of the most common treatment options for HCL and PLL are listed below. Treatment options and recommendations depend on several factors, including whether you have HCL or PLL, possible side effects, and your preferences and overall health. Your care plan may also include treatment for symptoms and side effects, an important part of cancer care. Take time to learn about all of your treatment options and be sure to ask questions about things that are unclear. Also, talk about the goals of each treatment with your doctor and what you can expect while receiving the treatment. Learn more about [making treatment decisions](#) [6].

Chemotherapy

Chemotherapy is the use of drugs to destroy cancer cells, usually by stopping the leukemia

cells? ability to grow and divide. Chemotherapy is given by a medical oncologist, a doctor who specializes in treating cancer with medication, or a hematologist, a doctor who specializes in treating blood disorders.

Systemic chemotherapy is delivered through the bloodstream to reach leukemia cells throughout the body. Common ways to give chemotherapy include an intravenous (IV) tube placed into a vein using a needle or in a pill or capsule that is swallowed (orally). A chemotherapy regimen (schedule) usually consists of a specific number of cycles given over a set period of time. A patient may receive one drug at a time or combinations of different drugs at the same time.

Drugs that are used to treat HCL include cladribine (Leustatin) and pentostatin (Nipent). For most people with HCL, both cladribine and pentostatin can bring a remission that lasts for many years and are the standard initial treatment for the disease.

PLL is treated with a combination of drugs, and several different regimens can be used. The specific regimen recommended for each patient is based on the type of chemotherapy the patient may have received previously and the possible side effects.

The side effects of chemotherapy depend on the individual and the dose used, but they can include fatigue, risk of infection, nausea and vomiting, hair loss, loss of appetite, and diarrhea. These side effects usually go away once treatment is finished.

Learn more about [chemotherapy](#) [7] and [preparing for treatment](#) [8]. The medications used to treat cancer are continually being evaluated. Talking with your doctor is often the best way to learn about the medications prescribed for you, their purpose, and their potential side effects or interactions with other medications. Learn more about your prescriptions by using [searchable drug databases](#) [9].

Immunotherapy

Immunotherapy, also called biologic therapy, is designed to boost the body's natural defenses to fight the cancer. It uses materials made either by the body or in a laboratory to improve, target, or restore immune system function. Immunotherapy for HCL includes recombinant interferon alpha (Alferon, Intron A, Roferon-A). Learn more about [immunotherapy](#) [10].

Targeted therapy

Targeted therapy is a treatment that targets the leukemia's specific genes, proteins, or the tissue environment that contributes to growth and survival. This type of treatment blocks the growth and spread of leukemia cells while limiting damage to healthy cells.

Recent studies show that not all cancers have the same targets. To find the most effective treatment, your doctor may run tests to identify the genes, proteins, and other factors involved in your leukemia. As a result, doctors can better match each patient with the most effective treatment whenever possible. In addition, many research studies are taking place now to find out more about specific molecular targets and new treatments directed at them. Learn more about [targeted treatments](#) [11].

A monoclonal antibody is a type of targeted therapy. It is directed against a specific protein in the leukemia cells, and it does not affect cells that don't have that protein. Rituximab (Rituxan) is an antibody directed against the surface protein CD20. BL22 immunotoxin is an antibody that is designed to attach to the surface protein CD22 and deliver a toxin to the leukemia cell. Both rituximab and BL22 have been used to treat HCL that did not go into remission after the initial treatment. However, BL22 is not approved by the U.S. Food and Drug Administration (FDA) for the treatment of PLL and is only available in [clinical trials](#) [3]. Alemtuzumab (Campath) is an antibody that targets the surface protein CD52 and has been used to treat PLL. Talk with your doctor about possible side effects for a specific medication and how they can be managed.

Surgery

Surgery to remove the spleen, which also makes white blood cells, is called a splenectomy and may be done for some patients. A surgical oncologist is a doctor who specializes in treating cancer using surgery. Learn more about [surgery](#) [12].

Stem cell transplantation/bone marrow transplantation

A stem cell transplant is a medical procedure in which bone marrow that contains leukemia is replaced by highly specialized cells, called hematopoietic stem cells, that develop into healthy bone marrow. Hematopoietic stem cells are blood-forming cells found both in the bloodstream and in the bone marrow. Today, this procedure is more commonly called a stem cell transplant, rather than bone marrow transplant, because it is the stem cells in the blood that are typically being transplanted, not the actual bone marrow tissue.

Before recommending transplantation, doctors will talk with the patient about the risks of this treatment and consider several other factors, such as the type of leukemia, results of any previous treatment, and patient's age and general health. Transplantation is a higher-risk procedure. It is not used often for people with HCL and PLL because it is not always an effective therapy for these diseases and because many patients with HCL or PLL are older and the risks of the procedure are higher.

There are two types of stem cell transplantation depending on the source of the replacement blood stem cells: allogeneic (ALLO) and autologous (AUTO). ALLO uses donated stem cells, while AUTO uses the patient's own stem cells. ALLO transplantation is sometimes considered for younger patients with PLL when chemotherapy was not effective. However, AUTO transplantation is rarely considered for patients with PLL or HCL.

In both types, the goal is to destroy all of the leukemia cells in the marrow, blood, and other parts of the body using high doses of chemotherapy and/or radiation therapy and then allow replacement blood stem cells to create healthy bone marrow. Learn more about [stem cell and bone marrow transplantation](#) [13].

Getting care for symptoms and side effects

HCL and PLL, as well as the treatments for these types of leukemia, often cause side effects. In addition to treatment to slow, stop, or eliminate the disease, an important part of care is relieving

a person's symptoms and side effects. This approach is called palliative or supportive care, and it includes supporting the patient with his or her physical, emotional, and social needs.

Palliative care can help a person at any stage of illness. People often receive treatment for the leukemia and treatment to ease side effects at the same time. In fact, patients who receive both often have less severe symptoms, better quality of life, and report they are more satisfied with treatment.

Palliative treatments vary widely and often include medication, nutritional changes, relaxation techniques, and other therapies. You may also receive palliative treatments similar to those meant to eliminate the leukemia, such as chemotherapy, surgery, and radiation therapy. Talk with your doctor about the goals of each treatment in your treatment plan.

Before treatment begins, talk with your health care team about the possible side effects of your specific treatment plan and supportive care options. And during and after treatment, be sure to tell your doctor or another health care team member if you are experiencing a problem so it is addressed as quickly as possible. Learn more about [palliative care](#) [14].

Refractory HCL and PLL

If HCL or PLL continue to worsen despite treatment, it is called refractory or resistant. Patients with this diagnosis are encouraged to talk with doctors who are experienced in treating refractory HCL or PLL, because there can be different opinions about the best treatment plan. Learn more about seeking a [second opinion](#) [15] before starting treatment, so you are comfortable with the treatment plan chosen. This discussion may include [clinical trials](#) [3] studying new treatments.

For HCL, your health care team may recommend a treatment plan that includes rituximab and/or BL22 immunotoxin. Supportive care will also be important to help relieve symptoms and side effects. In addition, it has recently been shown that almost all patients with HCL have a mutation in their leukemia cells in a gene called *BRAF*. A drug that stops a mutated *BRAF* gene from helping the leukemia grow and spread called vemurafenib (Zelboraf) has been successful for treating melanoma and a few patients with refractory HCL have received this drug. Talk with your doctor about your treatment options.

For most patients, a diagnosis of refractory HCL or PLL is very stressful and, at times, difficult to bear. Patients and their families are encouraged to talk about the way they are feeling with doctors, nurses, social workers, or other members of the health care team. It may also be helpful to talk with other patients, including through a support group.

Recurrence and the chance of remission

A remission is when HCL or PLL cannot be detected in the body and there are no symptoms. A remission can be temporary or permanent. This uncertainty leads to many survivors feeling worried or anxious that the disease will come back. While many remissions are permanent, it's important to talk with your doctor about the possibility of the leukemia returning. Understanding the risk of recurrence and the treatment options may help you feel more prepared if the disease does return. Learn more about [coping with the fear of recurrence](#) [16].

If HCL or PLL does return after the original treatment, it is called recurrent HCL or PLL. When this occurs, a cycle of testing will begin again to learn as much as possible about the recurrence. After testing is done, you and your doctor will talk about your treatment options. Often the treatment plan will include the therapies described above such as chemotherapy, immunotherapy, targeted therapy, and stem cell transplantation, but they may be used in a different combination or given at a different pace. For instance, cladribine and pentostatin can work again to treat HCL if it recurs after a long period of remission. Your doctor may also suggest clinical trials that are studying new ways to treat recurrent HCL or PLL.

People with recurrent HCL or PLL often experience emotions such as disbelief or fear. Patients are encouraged to talk with their health care team about these feelings and ask about support services to help them cope. Learn more about [dealing with a recurrence](#) [17].

If treatment fails

Recovery from HCL and PLL is not always possible. If treatment is not successful, the disease may be called advanced or terminal HCL or PLL.

This diagnosis is stressful, and this is difficult to discuss for many people. However, it is important to have open and honest conversations with your doctor and health care team to express your feelings, preferences, and concerns. The health care team is there to help, and many team members have special skills, experience, and knowledge to support patients and their families. Making sure a person is physically comfortable and free from pain is extremely important.

Patients who have advanced HCL or PLL and who are expected to live less than six months may want to consider a type of palliative care called hospice care. Hospice care is designed to provide the best possible quality of life for people who are near the end of life. You and your family are encouraged to think about where you would be most comfortable: at home, in the hospital, or in a hospice environment. Nursing care and special equipment can make staying at home a workable alternative for many families. Learn more about [advanced cancer care planning](#) [18].

After the death of a loved one, many people need support to help them cope with the loss. Learn more about [grief and loss](#) [19].

The next section helps explain clinical trials, which are research studies. Use the menu on the side of your screen to select About Clinical Trials, or you can select another section, to continue reading this guide.

Links:

[1] <http://www.cancer.net/cancer-types/leukemia-b-cell-prolymphocytic-leukemia-and-hairy-cell-leukemia/treatment-options>

[2] <http://www.cancer.net/about-us>

[3] <http://www.cancer.net/node/19086>

[4] <http://www.cancer.net/node/19089>

[5] <http://www.cancer.net/node/25356>

[6] <http://www.cancer.net/node/24582>

[7] <http://www.cancer.net/node/24723>

[8] <http://www.cancer.net/node/24473>

- [9] <http://www.cancer.net/node/25369>
- [10] <http://www.cancer.net/node/24726>
- [11] <http://www.cancer.net/node/24729>
- [12] <http://www.cancer.net/node/30689>
- [13] <http://www.cancer.net/node/24717>
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- [15] <http://www.cancer.net/node/25355>
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- [17] <http://www.cancer.net/node/25042>
- [18] <http://www.cancer.net/node/25113>
- [19] <http://www.cancer.net/node/25111>