

Hairy Cell
Leukemia
HCL



WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with hairy cell leukemia (HCL). What does it mean and how will it affect you?

This fact sheet will help you:

Learn about HCL
and how it is
diagnosed

Get an overview
of treatment
options

Understand
what happens
next

What is leukemia?

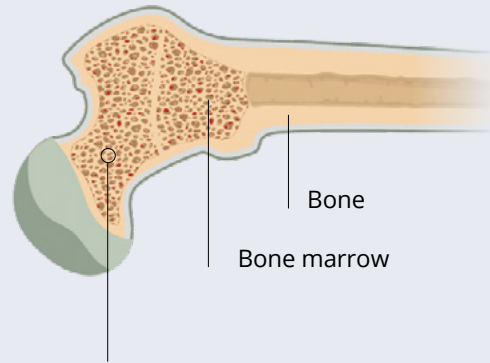
Leukemia is a cancer of the blood and bone marrow. Bone marrow is the soft, spongy material inside bones. Blood cells are formed in the bone marrow.

HCL is a rare type of leukemia in adults.

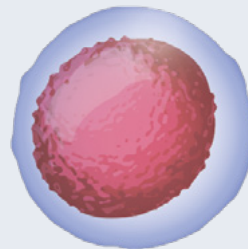
When you have leukemia, cancerous blood cells form and push out healthy blood cells.

Recent advances in treatments have made it possible to survive longer and enjoy a good quality of life for years.

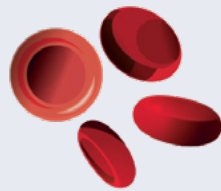
Blood is created in the **bone marrow** (the spongy part inside the bone).



Stem cell



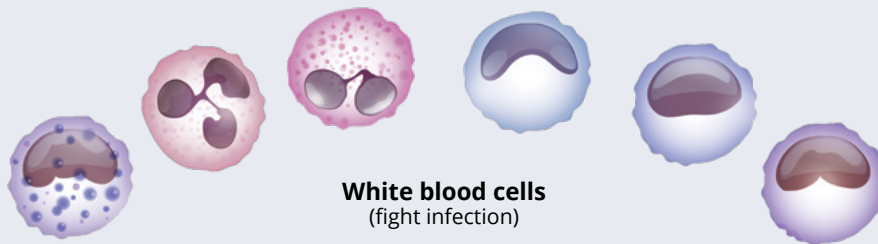
Three kinds of blood cells develop from stem cells:



Red blood cells
(carry oxygen)



Platelets
(allow blood to clot)



White blood cells
(fight infection)



About HCL

- Uncommon, slow-growing cancer
- Starts in a B-cell (type of white blood cell that is important for your immune system)
- Changes in the B-cell gene cause it to become malignant (harmful), grow and multiply uncontrollably
- These harmful cells enter the bone marrow and spleen and can attack your liver and lymph nodes
- The result is often a lower-than-normal number of healthy red and white blood cells and platelets, making you more prone to infections
- Chronic (long-lasting) type of leukemia; in most people, the cancer will return and will require more treatment
- Known as hairy cell because of the short, thin, hair-like projections on its cells

Risk factors

Certain factors can increase your risk of getting HCL:

- Your age (median age at diagnosis is around 58 years)
- Your gender (more common in men)
- Having a specific genetic mutation (change)
- Repeated exposure to chemicals, including certain herbicides

Signs and symptoms

Most people with HCL have symptoms related to an enlarged spleen or low blood cell counts. These include weakness and fatigue, infections, and/or bruising and bleeding. Combinations of these symptoms are common. Some people with HCL have no obvious signs or symptoms and the disease is uncovered during a routine blood test.

You may experience:

- Fatigue, weakness, shortness of breath during normal physical activities, and weight loss
 - When your red blood cell count is low (anemia)
- Bruising easily and ongoing bleeding from minor cuts
 - When your platelet count is low (thrombocytopenia)
- Infection
 - When your neutrophils and monocytes (types of white blood cells) count is low, your immune system is not working properly to guard against infection
- Abdominal pain or discomfort
 - When your spleen is enlarged and swollen

After your diagnosis

With your diagnosis, your doctor can determine the right treatment for you. Your test results help your doctor predict how HCL will likely progress and how you may respond to treatment. HCL can easily be confused with other blood diseases, so an accurate diagnosis is important to determine the best treatment options.

Name of test	Description
Medical history and physical exam	The doctor reviews past illnesses, injuries, and symptoms. They examine your lungs, heart, and other organs. They pay close attention to your lymph node areas (such as your head, neck, armpits, and groin).
Complete blood count (CBC) with differential	This test measures the number of red blood cells, white blood cells, and platelets in a sample of your blood. Often a person with HCL has low counts of each type. The “differential” measures the different types of white blood cells.
Peripheral blood smear	This test looks at blood cells under a microscope to see the number, size, shape, type, and pattern of cells. Cells that appear small- to medium-sized with the presence of hair-like projections are HCL cells.
Bone marrow aspiration and biopsy	These two tests look at bone marrow cells for anything unusual in your chromosomes. They are usually done at the same time.
Flow cytometry	During this test, cells are taken from your blood or tissue biopsy and then classified according to their cell surface proteins. The cell surface protein pattern of HCL cells is different from healthy B-cells and other types of abnormal B-cells.
Molecular testing	These very sensitive DNA (genetic) tests check for specific genetic changes in your cells. In most HCL cases, the leukemia cells have a mutation of a specific gene.
Imaging tests	A CT scan or ultrasound assesses your spleen, liver, and lymph nodes.



HCL treatment

HCL treatment can vary from one person to another. The start of your treatment will depend on your symptoms and whether your blood count is stable. The goal of treatment is to achieve a complete remission.

A complete remission means:

- Normal blood count level
- No hairy leukemia cells in your blood and bone marrow
- Reduction in the size of your spleen
- No other symptoms

Types of treatment

Watch and wait or active surveillance

delays treatment until the disease progresses, and signs and symptoms of HCL appear or get worse.

- This approach is for people with no symptoms at the time of diagnosis and a stable blood count.
- Some people with HCL live symptom-free for many years with this approach.

Chemotherapy

uses medicine (chemicals) to kill cancer cells. A combination chemotherapy procedure uses two or more chemotherapy drugs.

- With HCL, this treatment will start right away if you have blood counts below normal or have symptoms that include unexplained weight loss, infections that keep coming back, or physical discomfort from an enlarged spleen or liver.
- It is effective in 80% to 85% of people and can result in a complete remission that can last for many years.

Talk to your medical team about all treatment options, including clinical trials (new treatments being studied).

Factors that affect treatment

Discuss your treatment options with your doctor to make sure you understand the benefits and risks of each approach. Your treatment plan is based on:

- Your age and overall health status
- The type and extent of your symptoms
- Risk factors linked to a higher possibility that HCL will return after treatment:
 - Enlarged spleen
 - Hairy cells in your blood
 - Unmutated (unchanged) IGHV gene



Treatment side effects

When you begin your treatment for HCL, you may experience mild to severe side effects, depending on your age, your overall health, and your treatment plan. Most side effects disappear once your treatment ends. New drugs and therapies can help control most side effects. Speak to your doctor if you are having side effects.

Common side effects

You may experience side effects such as:

- Infection that can be severe from low white blood cell counts and low neutrophils (white blood cells that help fight infection)
- Nausea, diarrhea, vomiting, loss of appetite, aches and pains, low blood pressure, and temporary hair loss from chemotherapy
- Fatigue and shortness of breath from low blood counts and chemotherapy
- Fever, chills, infection, rash, mouth sores, and swelling from chemotherapy
- Low levels of red and white blood cells and platelets from chemotherapy

Long-term or late effects of treatment

Medical follow-up is important after treatment for HCL. You may need blood tests, bone marrow tests, or molecular tests to determine if you need further treatment. Your medical team should provide you with a care plan listing how often you will need follow-up visits and the tests you will have at those visits. If your blood count starts to drop, your doctors will discuss treatment options with you.

- **Long-term side effects** are common and can last for months or years after treatment ends. Examples include chronic fatigue and problems concentrating (known as chemo brain).
- **Late effects** are medical problems that do not show up until years after treatment ends. See your doctor to get follow-up care for possible early detection of heart disease and secondary cancers.





Living with HCL can be overwhelming. Seek medical help if you are feeling “down” or “blue” or don’t want to do anything and your mood does not improve over time. These could be signs of depression, an illness that should be treated even when you’re undergoing treatment for HCL. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

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This publication was made possible
thanks to the support of:



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