

What Is Hairy Cell Leukemia? Experts Explain This Rare Type of Cancer

An enlarged spleen can be a symptom, and chemotherapy is usually the go-to treatment.



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September 10, 2021

You probably know that "leukemia" means [blood cancer](#). You might even be familiar with some of the most common types, such as acute myeloid leukemia (AML) or [chronic lymphocytic leukemia](#) (CLL). But unless you or a loved one has been diagnosed with hairy cell leukemia (HCL), you've probably never heard of it.

That makes sense considering that this form of leukemia only accounts for about 2% of leukemia cases, according to the [National Organization for Rare Disorders](#). And there are only about 61,000 new cases of *any* kind of leukemia in the US each year, the [Cleveland Clinic](#) reports.

The fact that HCL is rare, however, probably doesn't matter much for people who are dealing with the diagnosis. Getting the facts is key, and a few basics might ease your fears: For starters, as the [American Cancer Society](#) (ACS) points out, HCL is a slow-growing (aka chronic) form of leukemia, and the current treatments for it usually work very well.

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What is hairy cell leukemia?

Leukemia is a broad umbrella term that refers to cancer of the blood and bone marrow (where blood cells are formed), [ACS](#) explains. While people with various types of leukemia make too many abnormal white blood cells, HCL specifically causes the overproduction of lymphocytes (aka B cells), which are a type of white blood cell that fights infection, says the [Mayo Clinic](#).

The unusual-sounding name comes thanks to the fact that the cancer cells look like they have tiny hairs protruding from them when they're viewed under a microscope, says Jeffrey Schriber, MD, director of Hematologic Malignancies at Cancer Treatment Centers of America in Phoenix, Arizona.

Who gets hairy cell leukemia?

HCL is more apt to develop in men who are middle aged or older, though some women and younger people do get it. It seems to be more prevalent among Ashkenazi Jews (of Eastern European origin) compared to people of other backgrounds, reports the [American Society of Clinical Oncology](#).

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Hairy cell leukemia symptoms

"Hairy cell leukemia behaves like a lymphoma, but it's considered leukemia because it's in the bone marrow," says Dr. Schriber. (Lymphoma, in contrast, mostly impacts the lymph nodes.) While many leukemia patients develop symptoms like extreme fatigue, random fevers, or start to bleed or bruise very easily, HCL usually causes different problems that are more reminiscent of lymphomas.

"[HCL] patients get a gigantic spleen, and they often get gastrointestinal symptoms," says Dr. Schriber. "They might be nauseated, get full very quickly when eating, or complain of constipation or diarrhea. You can often feel a big mass in the abdomen," which is where the spleen is.

Because there are often similarities between leukemia and lymphoma—and there are many varieties of each—it's advisable to go to a major cancer center so you can get a correct, specific diagnosis. "Once you get the diagnosis you can figure out how to move forward," says Dr. Schriber.

How hairy cell leukemia is diagnosed

As with other types of leukemia, diagnosing HCL starts with blood tests followed by a bone marrow aspiration and biopsy, says the [Leukemia & Lymphoma Society](#) (LLS). These samples will be sent to a lab and be analyzed by a pathologist. If you do in fact have HCL, the pathologist should be able to detect hairy cells in your blood. They'll also have to analyze the cells to see if there's a characteristic marker on the surface of the lymphocytes.

With most types of cancer, patients are assigned a stage (1-4) based on how far the cancer has metastasized (spread). But according to the [National Cancer Institute](#), HCL is different because it's already in the bloodstream and bone marrow. Instead, oncologists typically suggest a treatment regimen based on several factors. Per [LLS](#), current blood counts, whether you're carrying any genetic mutations, and whether your spleen is enlarged (because a significant number of cancer cells have accumulated there) are among the considerations.

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Hairy cell leukemia treatment

Given that HCL is rare you might be expecting treatment options to be limited or lackluster, but that's not the case. Most of the time, the prognosis is excellent, says Dr. Schriber. In fact, 5-year survival rate is about 90%, says [LLS](#).

When someone has a leukemia like HCL that makes them overproduce white blood cells, they usually fall short on red blood cells (which carry oxygen) and platelets (important for blood clotting), says [LLS](#). Healthy white blood cells (as opposed to defective/cancerous ones) also tend to be lacking, it adds. Because people with HCL overproduce lymphocytes, a type of white blood cell, they're usually treated with a specific chemotherapy that restores normal blood counts by targeting and killing off lymphocytes.

A chemotherapy drug called cladribine is usually the first-choice treatment, according to [LLS](#). About 85% of patients who try it go into complete remission, LLS notes. A similar chemo drug called pentostatin tends to be equally effective, Mayo Clinic points out.

Patients who don't respond sufficiently to either of these drugs are among those who might be good candidates for targeted therapy. Also called biologic therapy, these are drugs that target your own immune system and teach it to destroy the cancer. Targeted treatments for HCL include rituximab and interferon, per Mayo Clinic. These drugs are usually administered via infusion.

Most people with HCL don't need surgery, says LLS, since the above-mentioned drugs are so effective. But in some cases, spleen removal (splenectomy) —which years ago was the first-line treatment—might still be in order. You'll probably only need surgery if you haven't responded well to the various drug treatments that are now available or if your cancer keeps recurring.

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