CONDITION

Hairy cell leukemia

Hairy cell leukemia is a slow-growing cancer of infection-fighting white blood cells.

WHAT IT IS
Slow-growing cancer of B lymphocytes

BODY SYSTEM
Blood, bone marrow, and immune system

MAIN SYMPTOMS
Tiredness, repeated infections, easy bruising, abdominal fullness

WHAT CAUSES IT
Acquired cell changes, usually including a BRAF mutation

HOW IT IS DIAGNOSED
Blood flow cytometry and bone marrow biopsy

MAIN TREATMENT
Monitoring, chemotherapy, antibody treatment, or targeted therapy

CAN IT BE CURED?
No - treatment often brings long remissions

IS IT AN EMERGENCY?
Sometimes - fever, severe breathlessness, or uncontrolled bleeding

LONG-TERM OUTLOOK
Often long survival with treatment and ongoing monitoring

Overview

Hairy cell leukemia is a rare, usually slow-growing blood cancer. It begins in B lymphocytes, a type of white blood cell that normally helps protect you from infection.

The abnormal cells have fine, hair-like projections when viewed under a microscope. This appearance gives the condition its name. The cells are not related to hair, and hairy cell leukemia does not affect hair growth directly.

Abnormal cells can collect in the bone marrow, blood, spleen, and sometimes the liver. Bone marrow is the soft tissue inside certain bones where blood cells are made. As leukemia cells build up, there may be less room to produce healthy red blood cells, white blood cells, and platelets.

Low blood cell counts can cause tiredness, infections, bruising, or bleeding. An enlarged spleen

may cause discomfort or fullness under the ribs on the left side.

Hairy cell leukemia often develops slowly. Some people do not need treatment when it is first found. When treatment is needed, it is usually very effective and can control the condition for many years. However, the leukemia may return, so long-term follow-up remains important.

Symptoms

Some people have no symptoms. The condition may be discovered when a routine blood test shows low blood cell counts.

Possible symptoms include:

Anemia means there are too few healthy red blood cells to carry oxygen. Low neutrophils, a type of white blood cell, can make infections more likely. Low platelets can make bruising and bleeding easier.

These symptoms have many possible causes. Having one or more does not mean that you have hairy cell leukemia.

- Tiredness, weakness, breathlessness, or pale skin from anemia
- Frequent, severe, or slow-to-clear infections
- Easy bruising, nosebleeds, bleeding gums, or small red skin spots
- Fullness, discomfort, or pain under the left ribs from an enlarged spleen
- Reduced appetite or feeling full after eating a small amount
- Unexplained weight loss, fever, or night sweats

Causes and risk factors

Hairy cell leukemia develops when a B lymphocyte gains changes in its genetic material. These are acquired changes, meaning they develop during a person's lifetime rather than usually being inherited from a parent.

Most cases of classic hairy cell leukemia contain a change called the BRAF V600E mutation . This change keeps a growth signal inside the cell switched on. The abnormal cell can then survive and multiply when it should not.

Researchers do not know why this mutation develops. Hairy cell leukemia is more often diagnosed in older adults and in men, but it can affect other people as well.

There is no clear evidence that ordinary daily activities, diet, stress, or something you did

caused the condition. It is not contagious and cannot be passed between people.

A condition once called "hairy cell leukemia variant" is now generally recognized as a separate type of blood cancer. It often lacks the BRAF mutation and may respond differently to treatment. Accurate testing is important because several blood conditions can produce cells that look hairy under a microscope.

Diagnosis

Diagnosis usually begins with a full blood count. This test measures red blood cells, different types of white blood cells, and platelets. Hairy cell leukemia often causes low levels of more than one blood cell type.

A blood smear allows a laboratory specialist to examine the shape of the cells under a microscope. Hair-like projections may suggest the diagnosis, but appearance alone is not enough to confirm it.

Flow cytometry examines proteins on the surface of blood or bone marrow cells. Hairy cells have a characteristic combination of these proteins. This testing helps distinguish classic hairy cell leukemia from similar conditions.

A bone marrow biopsy is usually part of the assessment. A clinician removes a small core of bone marrow, usually from the back of the hip, using local anesthetic. The sample shows how much of the marrow contains leukemia cells and whether the tissue has become scarred. Scarring can sometimes make it difficult to withdraw liquid marrow, which is known as a "dry tap."

Testing for the BRAF V600E mutation can support the diagnosis and may help guide treatment if the condition returns. Imaging, such as an ultrasound or CT scan, may be used to assess an enlarged spleen or liver, but imaging does not confirm hairy cell leukemia by itself.

Treatment and management

Treatment is based on your symptoms, blood counts, spleen size, infections, general health, and previous treatment. Not everyone needs treatment immediately.

If you feel well and your blood counts are stable, active monitoring may be appropriate. This is sometimes called "watch and wait." It involves scheduled appointments and blood tests rather than ignoring the condition. Treatment can begin if meaningful changes develop.

Treatment is usually considered when low blood counts are causing problems, infections are

recurring, the spleen is uncomfortable or very enlarged, or other symptoms are affecting daily life.

The main medicines used for classic hairy cell leukemia include:

A purine analogue may be used alone or with an antibody treatment. The best approach can differ when there is an active infection, significant immune suppression, a previous relapse, or another medical condition.

Treatment can temporarily lower healthy blood cells before the marrow recovers. This may increase the risk of infection, anemia, or bleeding. Blood counts are therefore monitored during and after treatment. Recovery may take weeks or months, even when the leukemia has responded well.

If hairy cell leukemia returns, treatment options may include repeating a medicine that previously produced a long remission, adding antibody treatment, using targeted therapy, or joining a clinical trial. Removal of the spleen and treatment with interferon are now used less often, but may still have a role in selected situations.

Supportive care can include treatment for infections, blood or platelet transfusions, and advice about vaccination. Because some treatments weaken immune responses, vaccination plans should be discussed with the healthcare team managing the leukemia.

- Purine analogues , such as cladribine or pentostatin, which are chemotherapy medicines that damage leukemia cells
- Monoclonal antibodies , such as rituximab, which attach to a protein on B cells and help the immune system remove them
- Targeted medicines , including BRAF inhibitors, which block a growth signal used by leukemia cells with a BRAF mutation

Complications

Most complications result from low blood cell counts, an enlarged spleen, or the effects of treatment.

Too few neutrophils can lead to serious bacterial or fungal infections. Anemia may cause marked tiredness, dizziness, or breathlessness. Very low platelets can cause unusual bruising or bleeding.

A greatly enlarged spleen may cause ongoing discomfort, early fullness after meals, or, rarely, rupture after an injury. Sudden severe pain under the left ribs, especially after a fall or blow, needs emergency assessment.

Treatment can suppress the immune system for a time. Some infections may occur even after blood counts begin to improve. Your care team may arrange continued monitoring or preventive medicines based on the treatment you receive.

Studies have reported other cancers in some people with hairy cell leukemia. It is not always clear how much of this risk comes from the leukemia, its treatment, age, or other factors. Keeping up with recommended cancer screening and reporting new symptoms are sensible parts of follow-up.

Prevention

There is no established way to prevent hairy cell leukemia. The genetic changes involved are usually acquired by chance, and there is no proven lifestyle change that prevents them.

The condition is not normally inherited, so relatives do not usually need genetic testing or special screening simply because one family member has it.

Healthy habits can support general wellbeing, but they should not be presented as a way to prevent or cure hairy cell leukemia. During treatment, infection precautions and appropriate vaccines may reduce some complications. The advice needed will depend on your blood counts and immune function.

Living with it and what to expect

Hairy cell leukemia often follows a long-term, slow course. Many people have a strong response to their first treatment and remain in remission for years. Remission means there is no detectable or active leukemia causing problems; it does not necessarily mean every leukemia cell has gone.

Blood counts may recover gradually after treatment. Energy levels can also take time to improve, particularly if anemia or an infection was present. Some people return to usual work and activities quickly, while others need a slower recovery.

Follow-up usually includes discussion of symptoms, a physical examination, and blood tests. Bone marrow testing is not needed at every routine visit, but it may be used when the response is unclear or the condition appears to have returned.

Relapse is possible, sometimes many years later. A relapse does not automatically mean the outlook is poor. Further treatment can often produce another remission, although the choice may change if earlier remissions were short or the leukemia has become less responsive.

It is reasonable to ask how treatment may affect work, travel, sexual health, fertility, and vaccination. Infection risk may temporarily change what is safe, especially during periods of very low white blood cells.

When to get medical help

Contact your leukemia team or seek same-day medical advice if you develop:

Follow any fever threshold or emergency plan given to you during treatment. Infection can become serious more quickly when neutrophil levels are low.

Seek emergency help for severe difficulty breathing, chest pain, confusion, fainting, bleeding that will not stop, or sudden severe abdominal pain. Sudden pain under the left ribs after an injury could indicate damage to an enlarged spleen.

Routine follow-up is the right place to discuss gradual changes, ongoing fatigue, treatment side effects, or concerns about work and daily activities.

- Fever, chills, or signs of infection, especially during or after treatment
- New bruising, repeated nosebleeds, bleeding gums, or blood in urine or stool
- Worsening tiredness, dizziness, breathlessness, or reduced ability to exercise
- Increasing fullness or pain under the left ribs
- Symptoms that return after a period of remission

Common questions

Is hairy cell leukemia a type of cancer?

Yes. It is a cancer of B lymphocytes, which are infection-fighting white blood cells. It usually grows more slowly than many acute leukemias.

Why is it called "hairy" cell leukemia?

Under a microscope, the abnormal cells have thin projections around their edges that look like hairs. The name describes the cells' appearance and has nothing to do with body hair.

Can hairy cell leukemia be cured?

It is generally considered treatable rather than curable. Treatment often produces deep, long-lasting remissions, and many people live for years with little or no effect on everyday life. Continued monitoring is needed because the condition can return.

Will it always need treatment straight away?

No. If you have no significant symptoms and your blood counts are safe and stable, active monitoring may be recommended. Treatment is used when the condition begins causing important blood-count changes, infections, spleen symptoms, or other problems.

Is hairy cell leukemia inherited or contagious?

It is not contagious. The genetic changes in the leukemia cells are usually acquired during life, not inherited. Having the condition does not usually mean that your children or close relatives need screening.

Can hairy cell leukemia come back after treatment?

Yes. Relapse can happen, including after a long remission. Further treatment is often effective. The options depend on your previous response, current health, blood counts, and features of the leukemia cells.

Sources

- National Cancer Institute. Hairy Cell Leukemia Treatment (PDQ®) - Health Professional Version
- Grever MR, Abdel-Wahab O, Andritsos LA, et al. Consensus guidelines for the diagnosis and management of patients with classic hairy cell leukemia. Blood . 2017;129(5):553-560
- Jones G, Parry-Jones N, Wilkins B, Else M, Catovsky D. Guideline for diagnosis and management of hairy cell leukaemia and hairy cell variant. British Journal of Haematology . 2020;191(5):730-737
- Tiacci E, Trifonov V, Schiavoni G, et al. BRAF mutations in hairy-cell leukemia. New England Journal of Medicine . 2011;364:2305-2315