

Hairy cell leukaemia

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Quick overview of hairy cell leukaemia (HCL)

This section is an overview of the information on this page. There is more detail in the sections below.

What is it?

Hairy cell leukaemia is a type of cancer that develops from B lymphocytes (white blood cells that fight infection) and can affect the blood ('leukaemia'). It is slow-growing and usually responds very well to treatment. Most people diagnosed with hairy cell leukaemia are treated successfully and have a normal life expectancy.

What are the symptoms and how is it diagnosed?

The most common symptoms develop due to low blood counts, for example, recurring or long-lasting infections, tiredness, and easy bruising or bleeding. A swollen spleen (an organ of your immune system) can also cause abdominal (tummy) pain and a lump at the top-left of your abdomen.

Hairy cell leukaemia is diagnosed by looking at abnormal lymphocytes from blood tests and bone marrow tests.

How is it treated?

If you do not have any symptoms, you might be monitored ('active monitoring' or 'watch and wait') until you develop troublesome symptoms. Many people need to start treatment soon after diagnosis due to low blood counts. A short course of chemotherapy with cladribine or pentostatin is usually successful.

Some people need a second course of chemotherapy with or without rituximab (an antibody treatment) if the first course of treatment didn't clear the disease. If your spleen is very large, you might have an operation to remove it – a splenectomy. Hairy cell leukaemia commonly relapses (comes back), but this is often many years after first treatment. The same treatment options are usually successful for relapsed hairy cell leukaemia.

What is hairy cell leukaemia (HCL)?

Hairy cell leukaemia is a type of **cancer of the lymphatic system** that can develop when a lymphocyte (a type of white blood cell that fights infection) grows out of control. It is called a 'leukaemia' because it can be found in the blood and bone marrow (the spongy tissue in the centre of bones where blood cells are made).

It is called 'hairy cell' because the surface of the abnormal lymphocytes looks ruffled and hairy under a microscope.

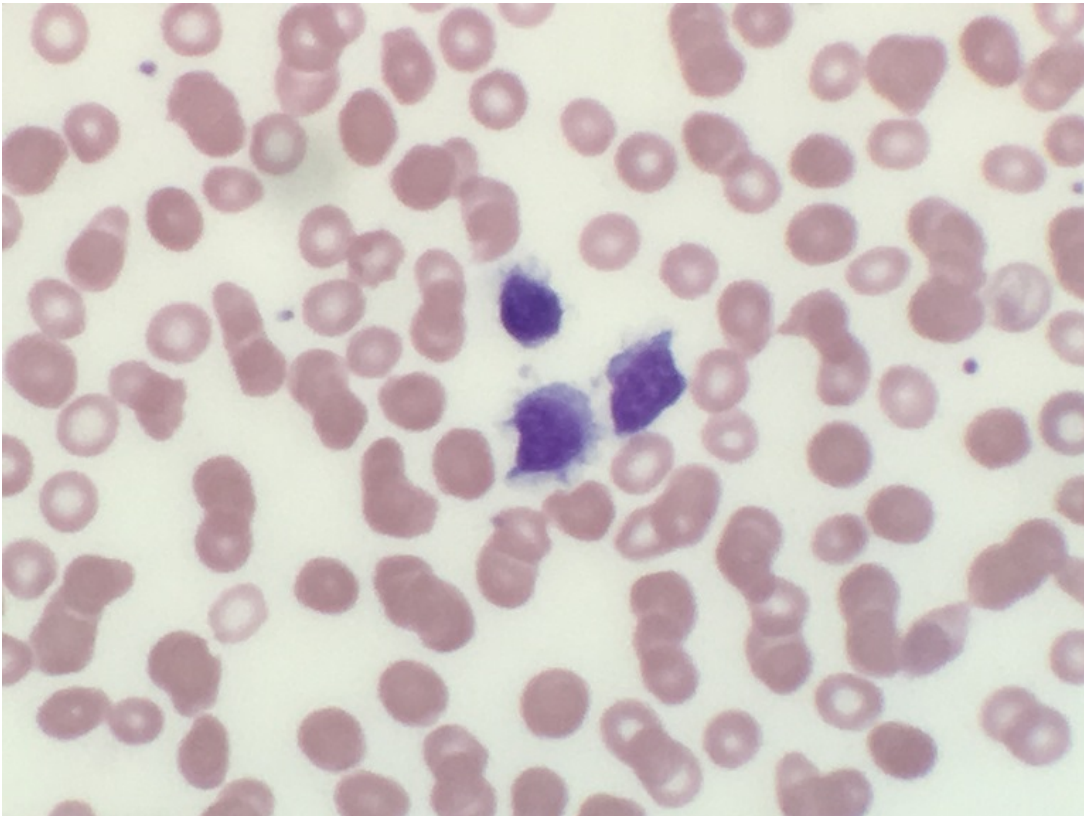


Figure: Hairy cell leukaemia cells stained purple

Who gets hairy cell leukaemia (HCL) and what causes it?

Hairy cell leukaemia is rare. Around 200 people are diagnosed with hairy cell leukaemia every year in the UK. Men are more likely to develop hairy cell leukaemia than women. It is most common in middle-aged and older people and it is very rare in young people.

There is no known **cause** of hairy cell leukaemia.

Symptoms

Some people have no **symptoms** when they are diagnosed with hairy cell leukaemia and it is found by chance on a blood test done for another reason.

The abnormal cells in hairy cell leukaemia are B lymphocytes that don't work properly. As the abnormal cells build up, the numbers of normal blood cells decrease and symptoms may develop. Some people have no symptoms at any point during their disease. The abnormal cells in hairy cell leukaemia grow slowly so any symptoms usually develop slowly. Most people have mild symptoms to start with and the symptoms gradually get worse as more abnormal cells build up.

You might feel generally unwell, with symptoms like fatigue (extreme tiredness), weight loss, fevers and night sweats. As your immune system is affected, you might have frequent infections or find it difficult to shake off infections. It is also quite common for abnormal cells to build up in your spleen (an organ of your **immune system**). This can make your spleen swell, which can cause pain or discomfort. You or your doctor might be able to feel a lump at the top-left of your abdomen (tummy) if your spleen is swollen. Your liver might also swell, which can also cause pain and swelling in your abdomen.

The abnormal cells usually build up in the bone marrow. This can cause **low blood counts** as the abnormal cells take up the space of normal cells.

It is common to have monocytopenia (low monocytes – a type of white blood cell), which can make you more prone to infection. Monocytopenia is a characteristic symptom of hairy cell leukaemia.

Other low blood counts that you might develop include:

- **anaemia (low red blood cells)**, which can cause tiredness and shortness of breath
 - **thrombocytopenia (low platelets)**, which makes you more likely to bruise and bleed
 - **neutropenia (low neutrophils – another type of white blood cell)**, which makes you more prone to infection.
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Diagnosis and staging

Hairy cell leukaemia is usually first found in **blood tests**. These typically show low blood counts together with abnormal lymphocytes.

If your doctors suspect that you have hairy cell leukaemia, you are likely to have **bone marrow tests** to see if there are abnormal cells in your bone marrow.

Experts look at the abnormal lymphocytes from your blood and bone marrow in more detail and do tests on them to make a diagnosis.

You have other tests, too, for example:

- more blood tests to find out about your general health
- a physical examination to check for other signs of hairy cell leukaemia such as an enlarged (swollen) spleen.

You might have a **scan**, usually a CT scan or **ultrasound scan**, to see if your spleen is enlarged or you have any large lymph nodes (glands) in your abdomen. If any large lymph nodes are found, a sample of them might be taken (a **biopsy**) to check whether the hairy cell leukaemia has **transformed** (changed) into a faster-growing type of lymphoma. This is a very rare complication of hairy cell leukaemia and needs different treatment.

Although **waiting for the results of your tests** can be difficult, your specialist is collecting important information during this time. It is important that your specialist knows exactly what type of cancer you have and how it is affecting you so they can give you the most appropriate treatment.

Hairy cell leukaemia is not usually given a '**stage**' (the stage refers to the extent of the cancer) as many of the other types of lymphoma are. Your medical team look at the results of your tests and other factors like your general health to decide whether or not you need treatment straightaway and what the best treatment is for you.

Outlook

The outlook for hairy cell leukaemia is usually very good. Most people with hairy cell leukaemia have a normal life expectancy. Treatment usually puts the disease into remission (no evidence of hairy cell leukaemia in your tests) and remissions often last many years. Hairy cell leukaemia can **relapse (come back)** but nearly all people who relapse are treated successfully again.

Your specialist is best placed to advise you on your outlook based on your individual circumstances. They can use the results of your tests and consider other factors, like your age, symptoms, and other conditions you have to predict how likely you are to respond well to a particular treatment. These factors are called 'risk factors'.

Treatment

If your hairy cell leukaemia is not yet causing problems, you might not need treatment straightaway. You might be monitored regularly by your specialist until you need treatment ('**active monitoring**' or '**watch and wait**'). Research shows that there is no benefit to starting treatment before you need it. All treatment also has a risk of causing side effects so specialists often prefer to delay treatment for as long as possible.

Most people diagnosed with hairy cell leukaemia have low blood counts and/or a swollen spleen. If you have these problems, you are more likely to start treatment soon after you are diagnosed.

Most people have **chemotherapy** as a first treatment for hairy cell leukaemia. You may also have other treatments to help support your body

including:

- **granulocyte-colony stimulating factor** (G-CSF) to boost your number of white blood cells, which fight infection
- **erythropoietin** to boost your red blood cells, which carry oxygen around your body.

A few people might be given a drug called '**interferon**' before starting chemotherapy. Interferon is given to treat the leukaemia in a chemotherapy-free way to boost your blood counts. Interferon is given three times a week by subcutaneous injection (injection under the skin) and you continue to have treatment until your blood counts are considered high enough for you to start chemotherapy. The main **side effects** of interferon are flu-like symptoms and muscle aches. Let your medical team know if you have any side effects as there are often other treatments that can help.

You are also likely to have treatment for any active infections before you start chemotherapy as chemotherapy reduces your immune system further.

Occasionally, your medical team might suggest a **splenectomy** (an operation to remove your spleen) before your chemotherapy. This is usually considered only if your spleen is very enlarged and causing serious problems. A few people with hairy cell leukaemia do not need any more treatment after having a splenectomy so you may be monitored for several months to see if further treatment is needed. Most people start chemotherapy several months after their splenectomy.

Chemotherapy for hairy cell leukaemia

The most common **chemotherapy** drugs are:

- **cladribine**, which is given every day for 5–7 days. It is usually given by subcutaneous injection (injection under the skin).
- **pentostatin**, which is given intravenously (into a vein) once every two weeks. It is given until most of your blood counts return to normal levels, which is usually around 3–4 months after starting treatment.

Both drugs are very effective on their own for hairy cell leukaemia. You can discuss the treatment options with their doctor to decide on the best course

of treatment for you.

You have frequent blood tests during and after your treatment. You then have tests to check your response to treatment once your blood counts return to normal. If you have cladribine, you have tests to check that you are clear of hairy cell leukaemia 3–6 months after finishing treatment. If you have pentostatin, you have these tests at the end of treatment.

Most people have a complete response to treatment (no hairy cell leukaemia left in your tests). People who have a complete response generally have a longer first remission than those who have a partial response to treatment. A partial response is when hairy cell leukaemia is reduced but there is still some disease remaining after treatment. However, many people with a partial response to treatment still have a first remission that lasts several years.

Sometimes, specialists suggest another course of chemotherapy to improve the response for people who don't have a complete response. Hairy cell leukaemia is usually cleared by chemotherapy alone but your specialist might suggest adding the antibody treatment **rituximab** if you need a second course of treatment. However, it is also common practice for specialists to recommend waiting until the disease relapses (becomes more active again) before having more treatment.

Rituximab is usually given intravenously for 6–8 doses in total. The treatment schedule varies. It might be given weekly or once every 2 weeks and can be given alongside your other treatment or after your other treatment has finished.

Side effects of chemotherapy

All treatment has a risk of **side effects** (unwanted effects). Your medical team can give you more information about the typical side effects of the treatment they recommend for you.

Both cladribine and pentostatin increase your risk of developing infections so you are given other treatments to prevent serious infections during and after your chemotherapy. These treatments usually include antibiotics and anti-viral drugs.

Other common side effects of both treatments include:

- nausea (feeling sick)
- fever (a temperature above 38°C)
- skin rashes.

Pentostatin can also cause you to be more sensitive than usual to sunlight – you might hear this called ‘photosensitivity’. Your skin may burn more easily than usual so it is important to protect yourself from the sun during treatment and for several months after.

You can be given treatments to help deal with any side effects.

Macmillan Cancer Support have more information about [cladribine](#) and [pentostatin](#) and the side effects associated with these treatments. We have more information about [rituximab](#), including information on side effects.

Note: after you have had treatment with cladribine or pentostatin you must be given specially prepared ‘irradiated blood’ if you need to have [blood transfusions](#) in future. This is so that you don’t develop a rare but serious complication of blood transfusion called ‘transfusion-associated graft-versus-host disease’.

Follow-up

When your hairy cell leukaemia is in remission, you have regular appointments at the hospital. This is called ‘[follow-up](#)’. Follow-up appointments are usually every 3–12 months but this varies between people depending on their individual circumstances. Some people have all their follow-up at the hospital. Some people who have normal blood counts and are well might prefer to be followed up by their GP and can be referred back to the hospital if their blood counts decrease or other symptoms develop. Follow-up appointments allow your medical team to check for signs of the lymphoma starting to [relapse](#). Even when hairy cell leukaemia is relapsing, any changes usually develop slowly.

Follow-up appointments also give you an opportunity to ask questions and raise any concerns.

You are likely to have a physical examination and blood tests. Scans are not usually done unless there is a particular reason for them.

If you stay well, your appointments may become less frequent.

If you are worried about your health at any time, contact your GP or medical team. They can reassure you or arrange an appointment for you to have a check-up.

Relapse

The first treatment for hairy cell leukaemia is often successful and many people remain in remission. However, it is common for hairy cell leukaemia to **relapse** (come back) and relapses can happen many years after your first treatment. Relapsed hairy cell leukaemia is usually treated successfully with the same chemotherapy drugs that are used as first treatments.

- If you relapse more than 2 years after your first treatment, you are likely to have the same treatment as you had before.
- If you relapse less than 2 years after your first treatment, the other chemotherapy drug is likely to be used. For example, if you had cladribine as a first treatment, pentostatin is likely to be used at relapse.

The antibody treatment rituximab might be added to your chemotherapy if you have relapsed. Nearly all people who relapse are treated successfully again with a second course of chemotherapy with or without rituximab.

Other treatment options might be considered, such as:

- **newer, targeted drugs**, possibly through a clinical trial
 - an **allogeneic stem cell transplant** if you are fit enough to have one and your medical team thinks your hairy cell leukaemia is likely to relapse again.
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Research and targeted treatments

Standard treatments are usually very successful for hairy cell leukaemia. Clinical trials continue to test new treatments for this type of lymphoma, most often for people with hairy cell leukaemia that is difficult-to-treat. Some studies have shown that newer drugs used in other types of low-grade non-Hodgkin lymphoma and other cancers could be effective for hairy cell leukaemia, for example **ibrutinib** and vemurafenib.

Our clinical trials information service, Lymphoma TrialsLink, has more information about clinical trials. You can search the database to find clinical trials currently open in the UK for people with hairy cell leukaemia.

Hairy cell leukaemia (HCL) variant

Hairy cell leukaemia variant (HCL-V) can cause similar symptoms to hairy cell leukaemia and the cells also look 'hairy' under a microscope. However, HCL-V is not considered to be related to hairy cell leukaemia. Like hairy cell leukaemia, HCL-V commonly causes low blood counts and a swollen spleen but it doesn't usually cause **monocytopenia**.

HCL-V can be more difficult to treat than hairy cell leukaemia. **Rituximab** is usually recommended, often with cladribine or after a **splenectomy**. Other treatments might be considered, particularly if HCL-V doesn't respond to the first treatment. Other treatments might include **targeted drugs**, a splenectomy if you haven't already had one or a **stem cell transplant**.

References

These are some of the sources we used to prepare this information. The full list of sources is available on request. Please contact us by email at **publications@lymphoma-action.org.uk** or phone on **01296 619409** if you would like a copy.

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Further reading

- [Glossary](#)
- [Splenectomy \(having your spleen removed\)](#)

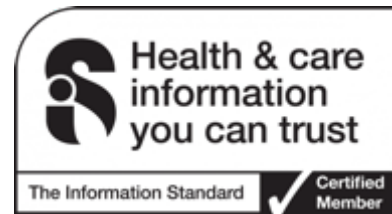
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