

# Hairy cell leukaemia (HCL)

**This fact sheet explains what hairy cell leukaemia (HCL) is and how it's diagnosed and treated. It explains the different types of HCL and suggests places to go for more information.**

## What is HCL?

Hairy cell leukaemia (HCL) is a rare form of leukaemia that's part of a family of related conditions called lymphoproliferative disorders (LPDs). Chronic LPDs are slow-growing blood cancers that affect a type of white blood cell called lymphocytes, which fight infections.

In HCL, these lymphocytes become abnormal and can no longer fight infections properly. These are also known as leukaemia cells. The condition gets its name because these cells have a very characteristic appearance: with hair-like outgrowths on the cell surface when it's examined under the microscope.

## Who gets HCL?

- › HCL is rare: about 100-200 people will get HCL per year in the UK.
- › 4 in 5 people who get HCL are male.
- › The average age of diagnosis is 57 years.
- › Children and teenagers don't get HCL.

## What causes HCL?

We don't know what causes HCL. It can't be passed from person to person and it doesn't run in families. It's recently been discovered that almost all people with HCL have a genetic change called BRAF that happens within the hairy cells, but not in the other cells in their body. This genetic change is clearly important in the development of HCL, but we don't yet know how or why this happens.

## Signs and symptoms

As HCL tends to be a slow-growing disease, some people are diagnosed by chance following a routine blood test for something else. However, most people with HCL will have some signs or symptoms before they're diagnosed. The most common ones include:

- › tiredness (fatigue), weakness and breathing problems, shortness of breath and palpitations, caused by a lack of red blood cells (a condition called anaemia),
- › bleeding and bruising, caused by a low number of platelets (cells that help to form blood clots),
- › discomfort in your upper stomach, caused by the swelling of your spleen because of the hairy cells within it,
- › frequent infections because you have fewer healthy white cells (the cells that fight infection),
- › low blood counts on your blood tests (known as pancytopenia), which is the result of too many abnormal cells being produced in your bone marrow.

## Diagnosis

HCL is diagnosed through blood and bone marrow tests. Your samples will be examined under a microscope in a laboratory.

Your doctors may suspect you have HCL from a blood sample as this can show hairy cells and a lower number of normal cells. A small number of people with HCL will have very few hairy cells in the blood, but they'll be in the bone marrow instead. Because of this, everyone will have a bone marrow examination (biopsy from your hip bone – from one side of the small of your back) to confirm your diagnosis.

You don't need to stay overnight in hospital for a bone marrow biopsy – you can have it as an outpatient using local anaesthetic or mild sedation. It's usually quite quick but will be uncomfortable while the sample is being taken from the marrow; you can take painkillers if you need to.

You can find more information about bone marrow tests on our website [bloodwise.org.uk](http://bloodwise.org.uk)

## Treatment

### Watch and wait

Your doctor may recommend that you don't need treatment straight away, if you're well and your blood count is relatively normal. You may just have regular check-ups and careful monitoring of your condition – this is called 'watch and wait'. However, for most people with HCL, it's important to treat the condition before your blood counts get too low.

### Chemotherapy

Most people with HCL will be offered treatment with chemotherapy drugs, either pentostatin or cladribine (cladribine is also called 2-CDA). These drugs are called purine analogues (PAs). They act by blocking the action of DNA in the hairy cells. There are no studies to show which drug is better, but strong evidence suggests that both drugs are equally effective.

Both of these drugs can be given into a vein (intravenous or IV). Cladribine can also be given as an injection under your skin (subcutaneous). The amount of time you'll be having chemotherapy will depend on the drug used and your treatment plan (drug regimen).

Subcutaneous cladribine is usually given as a course of one injection per day over five days. Pentostatin may be given by IV injection over a period of weeks or months. There's no evidence to suggest any one way of receiving the drugs is more or less effective than others.

PAs usually have few serious side effects, but you may get some mild side effects. Common side effects can include: feeling or being sick (nausea), rashes, extreme tiredness (fatigue), headaches and diarrhoea.

PAs can also lower your normal blood cell count further at first, and this can lead to infections which may mean you need to stay in hospital to have treatment with IV antibiotics. However, recovery is usually fast (three to six weeks).

Do let your healthcare team know if you have any side effects as often they'll be able to help you manage them – there are medicines you can take to help with nausea and vomiting. You'll also receive antibiotics to lower your risk of infections.

The electronic Medicines Compendium website has more information on side effects. Go to [medicines.org.uk/emc](http://medicines.org.uk/emc) and search for 'pentostatin' or 'cladribine'.

The drug you'll be given depends on your wishes (how you'd prefer the drugs to be given) and what your specialist suggests would be best for you.

### Interferon

A few people may be given a drug called interferon. This is the drug that was used to treat HCL before it was replaced by the PAs. Interferon is now mainly used for people who have very low levels of neutrophils (the cells that fight bacterial infections) and platelets (the cells that help to form blood clots). This is because the PAs can lower these cell counts further. Once your neutrophil and platelet counts return to a high enough level, you'll normally switch to a PA.

### Splenectomy

In some rare cases, people might need an operation to remove their spleen (splenectomy). These operations are rare because PAs are very effective at shrinking the spleen.

A splenectomy may benefit you if you have a very enlarged spleen but only a small amount of hairy cells (leukaemia cells) in your bone marrow. It's not a cure for the disease, but for a few people (about 5%) it may delay the need for drug treatment, possibly for many years. A splenectomy will also relieve the discomfort you get if your spleen is enlarged.

If you've had your spleen removed, you'll be at higher risk of certain types of serious infections. The risk isn't so great that you'd need to change your lifestyle, but you should see a doctor immediately if you feel ill, and tell them that you've had a splenectomy.

### Remission

After finishing treatment, your bone marrow will be examined again to see if all the abnormal hairy cells have disappeared. It's called a complete remission when there's no evidence of hairy cells in the bone marrow.

### Further treatment

You may be offered more treatment if your bone marrow tests show that some hairy cells are left. After you're treated with a PA, you usually stay in remission for a long time (an average of 15 years).

If the disease does return (known as a relapse) there's a very high chance that any further treatment you receive will be successful. This may involve taking the other purine analogue or re-using the same drug. You may also be given other drugs called monoclonal antibodies. These drugs are targeted therapies that can recognise specific proteins on the leukaemia cells and kill them. A monoclonal antibody called rituximab may be used.

There are other new targeted therapies now available, including a drug that targets and blocks the BRAF protein, called vemurafenib. However, these drugs may only be available in a clinical trial. Your specialist will discuss treatment options with you if you experience a relapse.

You can find out more information about clinical trials in our booklet **Clinical trials**.

### Follow up

You'll be monitored closely usually for the first two years after treatment by your consultant and GP. This will involve blood tests every few months. After this time, tests are done less often, usually every 6–12 months. It's important to let your doctor know if you feel unwell at any time while you're in remission.

### Outlook

Although the outlook for patients with HCL is excellent, there currently isn't a cure. The very effective drug treatments now available mean that most people diagnosed with HCL will have a normal or near-normal life expectancy. Short courses of treatment have few serious side effects. Most people will recover quickly following treatment and can return to a normal life.

## Finding out more



We offer patient information on many blood cancer types and topics, online and in free printed booklets. They cover everything from symptoms and diagnosis through to treatment and living with your condition. Go to [bloodwise.org.uk/information-and-support](https://bloodwise.org.uk/information-and-support)



We also have an online community you may like to join [bloodwise.org.uk/our-community](https://bloodwise.org.uk/our-community)



Or you can call our support line on **0808 2080 888** (Mon–Fri 10am–4pm). This is a freephone number.



See our website for more details of cancer information specialists and support groups. Go to [bloodwise.org.uk/living/where-get-help-and-support](https://bloodwise.org.uk/living/where-get-help-and-support)

## About Bloodwise

We're the UK's specialist blood cancer charity.

We've been working to beat blood cancer since 1960.

We fund world-class research; provide practical and emotional support to patients and their loved ones; and raise awareness of blood cancer.

We'd like to thank Dr Claire Dearden and Dr Edward Kanfer, for their help and support in developing the content and checking for clinical accuracy. A list of references used in this fact sheet is available on request, please email at [information@bloodwise.org.uk](mailto:information@bloodwise.org.uk).

Our factsheets contain general information. Always listen to the advice of your specialist about your individual condition – because every person is different.

### Disclaimer

We make every effort to make sure that the information in this fact sheet is accurate, but you shouldn't rely on it instead of a fully trained clinician. It's important to always listen to your specialist and seek advice if you have any concerns or questions about your health.

Bloodwise can't accept any loss or damage resulting from any inaccuracy in this information, or from external information that we link to.

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Bloodwise, 39–40 Eagle Street, London WC1R 4TH  
**020 7504 2200** | [info@bloodwise.org.uk](mailto:info@bloodwise.org.uk) | [bloodwise.org.uk](https://bloodwise.org.uk)

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