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Hairy cell leukaemia

This information is about hairy cell leukaemia, a slow-growing type of blood cancer. It also covers hairy cell leukaemia variant, which is a type of non-Hodgkin lymphoma.

On this page

We have separate information about the topics in **bold font**. Please get in touch if you'd like to request copies or if you would like further information about any aspect of lymphoma. Phone 0808 808 5555 or email **information@lymphoma-action.org.uk**.

What is hairy cell leukaemia?

Hairy cell leukaemia is a slow-growing type of blood cancer. It develops when white blood cells called **lymphocytes** grow out of control. Lymphocytes are part of your **immune system**. They travel around your body in your blood and **lymphatic system**, helping you fight infections. There are two types of lymphocyte: **T lymphocytes (T cells)** and **B lymphocytes (B cells)**. Hairy cell leukaemia develops from B cells.

In hairy cell leukaemia, the abnormal B cells build up in your blood and **bone marrow**. This is why it's called 'leukaemia' – after 'leucocytes': the medical name for white blood cells. Occasionally, the abnormal B cells can build up in your **lymph nodes**.

The abnormal B cells in hairy cell leukaemia look ruffled and hairy under a microscope. This is how it gets its name.

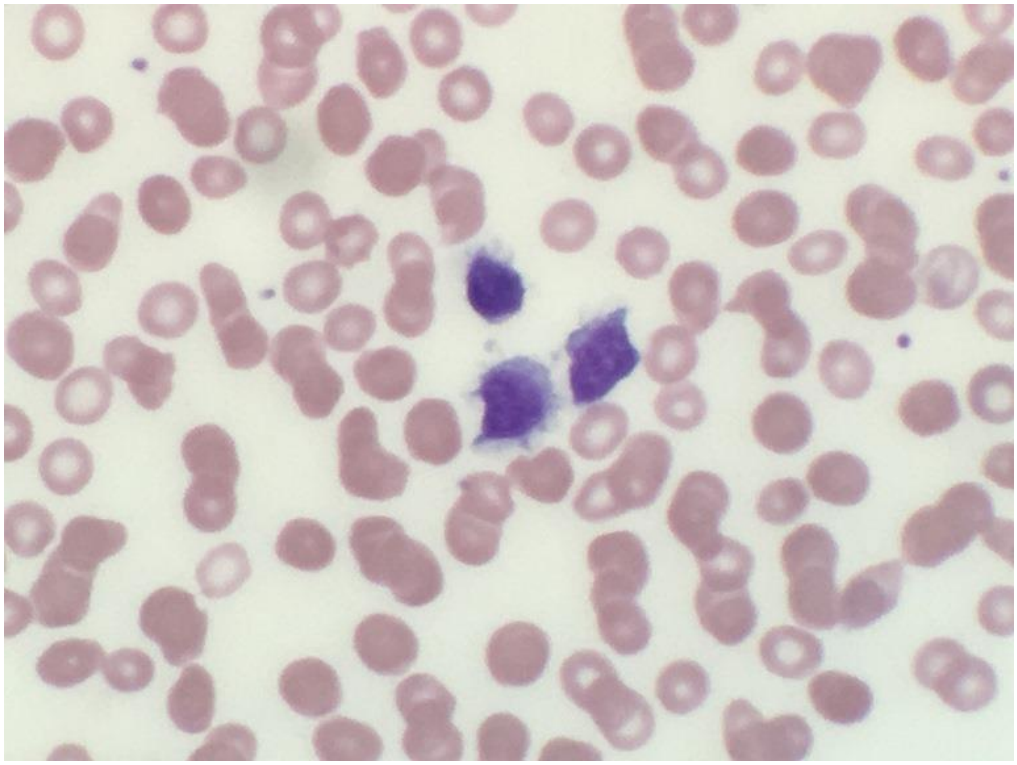

Figure: Hairy cell leukaemia cells (shown in purple)

Who gets hairy cell leukaemia?

Hairy cell leukaemia is rare. Only around 240 people are diagnosed with it each year in the UK.

Hairy cell leukaemia affects around four times more men than women. It typically develops in middle-aged people. It is 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 when a blood test is done for another reason.

Most people develop symptoms slowly, as the abnormal cells in hairy cell leukaemia grow. It is common for these cells to build up in your **bone marrow**, where they take up the space needed for healthy blood cells to develop. This means your body might not be able to make enough blood cells and you might develop low blood counts:

- **anaemia** (low red blood cells), which can make you feel tired, breathless or dizzy
- **thrombocytopenia** (low platelets), which makes you more likely to bruise and bleed
- **neutropenia** (low neutrophils – a type of white blood cell), which might make you get infections more easily than usual, and can make it difficult to get rid of them
- monocytopenia (low monocytes – another type of white blood cell), which affects almost all people with hairy cell leukaemia and can increase your risk of developing 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, discomfort, or a feeling of fullness in your tummy (abdomen). You or your doctor might be able to feel your swollen spleen as a lump at the top-left of your tummy. A very large spleen can also destroy healthy blood cells inside it, making your blood counts even lower. Your liver might also swell, which can cause pain and bloating. A few people get swollen nodes inside their tummy.

You might feel generally unwell, with symptoms like fatigue (extreme tiredness), weight loss, fevers and night sweats.

Very rarely, people with hairy cell leukaemia develop swollen lymph nodes without having abnormal cells in their blood, spleen or bone marrow. This can look very similar to some types of slow-growing (low-grade) non-Hodgkin lymphoma.

Diagnosis and staging

Hairy cell leukaemia is usually diagnosed on **blood tests**. These typically show low blood counts together with abnormal ‘hairy’ lymphocytes in your bloodstream. Your doctor will examine you to check for other signs of hairy cell leukaemia such as a swollen **spleen** or liver.

You are likely to have a **bone marrow biopsy** to check for abnormal cells in your bone marrow. An expert lymphoma **pathologist** looks at your blood and biopsy samples under a microscope. They run specialised tests on the abnormal cells to find out if they have certain genetic changes or make particular proteins.

You will have other blood tests to check your general health. If you are starting treatment, you might have blood tests to check if you have any viral infections that could flare up during treatment.

You might have a scan, such as a **CT scan** or **ultrasound scan**, to see if your spleen is enlarged or if you have any swollen lymph nodes in your tummy (abdomen). However, a scan isn’t always necessary because these can often be detected on a physical examination.

Very rarely, you might be diagnosed with hairy cell leukaemia based on a **biopsy** of your spleen or after having your spleen removed.

You usually have your tests done as an outpatient. It takes a few weeks to get all the results. **Waiting for test results** can be a worrying time, but it is important for your medical team to have all the information they need so they can plan the most appropriate **treatment** for you.

With most types of cancer, you are given a **stage** when you are first diagnosed. This tells you how widespread the cancer is in your body. Staging helps your medical team decide on the most appropriate treatment for you. For hairy cell leukaemia, there is no widely agreed staging system. Instead, your medical team use the results of your blood tests, physical examination and symptoms 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 is very successful and usually puts the disease into remission (no evidence of hairy cell leukaemia on your tests). Remissions often last many years but hairy cell leukaemia usually comes back (**relapses**) and needs more treatment at some point. 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.

Many people do not find exact outcome statistics helpful. They don't tell you what your individual outlook is – they only tell you how a group of people with the same diagnosis did over a period of time. They are usually measured over 5 or 10 years after treatment, which means they only tell you how people did in the past. Treatments are improving all the time, and those people might not have received the same treatment as you. However, if you do want to know more about survival statistics for hairy cell leukaemia, **Cancer Research UK** have some information that you might find useful.

Treatment

The treatment you have for hairy cell leukaemia depends on how it is affecting you.

Active monitoring

You might not need treatment straightaway if you don't have troublesome symptoms, your liver or spleen are not too swollen, and your blood counts aren't too low. Instead, your medical team monitors you every 3 to 6 months until you need treatment. This is called **active monitoring or watch and wait**.

If you are worried about your health at any time, contact your GP or medical team. You don't have to wait for your next appointment.

You are likely to start **treatment** if:

- you develop symptoms such as fever, night sweats or fatigue
- your spleen, liver or lymph nodes become very swollen
- your blood counts become too low

- you have lots of infections or severe infections.

Treatment options

Most people diagnosed with hairy cell leukaemia start treatment soon after diagnosis, but some people might not need treatment for months or even years.

Most people have **chemotherapy** as a first treatment. The most common chemotherapy drugs for hairy cell leukaemia are:

- **Cladribine**, which you have as an injection under your skin (subcutaneous injection) or through a drip into a vein (an intravenous infusion). Depending on how you are having cladribine and the usual practice at your hospital, you either have treatment every day for 5 to 7 days, or once a week for 5 to 6 weeks.
- **Pentostatin**, which you have through a drip into a vein once every two weeks. You carry on having treatment until most of your blood counts return to normal levels, which is typically 4 to 5 months after starting treatment.

Very rarely, your medical team might recommend having an operation to remove your spleen (a **splenectomy**). This is usually considered only if your spleen is very enlarged and causing serious problems.

Your medical team will discuss the options with you to decide on the best course of treatment for you. They will also give you information about the typical **side effects** of the treatment they recommend. Macmillan Cancer Support have more information about **cladribine**, **pentostatin** and **interferon-alfa** and their possible side effects.

You usually have **blood tests** every week during and after treatment to check your blood counts. Around 4 to 6 months after having cladribine, or when you finish pentostatin treatment, you are likely to have a **bone marrow biopsy**.

Your medical team uses the results of these tests to check how well your hairy cell leukaemia has responded to treatment.

- A complete response means your blood counts have returned to normal, there are no abnormal cells in your bloodstream or bone marrow, and you no longer have a swollen liver or spleen.

- A partial response means your blood counts have returned to normal, there are no abnormal cells in your bloodstream, your spleen and liver have shrunk by at least half, and the number of abnormal cells in your bone marrow has reduced by at least half.

Most people have a complete response to treatment. If you have a partial response to treatment, you might have another course of chemotherapy, sometimes combined with the antibody therapy **rituximab**. This increases your chance of having a long-lasting remission. Occasionally, hairy cell leukaemia doesn't respond well to your first treatment and you need more treatment straightaway.

Treatment to relieve or prevent symptoms

As well as treatments to control the hairy cell leukaemia, you are likely to have treatments to help relieve your symptoms or prevent infections. These might include:

- antibiotics and antiviral drugs to prevent or treat infections
- annual vaccinations against **flu** and **pneumonia**
- **red blood cell transfusions or platelet transfusions** to treat low blood counts (these have to be with specially treated [**irradiated**] blood products if you've had cladribine or pentostatin treatment, to prevent a reaction called 'graft-versus-host disease')
- **growth factor (G-CSF) injections** to boost your white blood cell count if you have an infection.

Contact your medical team straightaway if you have any **symptoms of infection**. It is important that you get prompt treatment.

Follow-up

When your hairy cell leukaemia is in **remission** after treatment, you have regular **follow-up appointments**. These might be at your hospital or at your local GP surgery. They are usually every 3 to 12 months.

At your appointment, you have blood tests and your doctor or nurse specialist examines you and asks if you have any concerns or symptoms. Your follow-up appointments are to check:

- how well you are recovering from treatment
- for any signs that the hairy cell leukaemia might be relapsing

- that you are not developing any **late effects** (side effects that can develop months or years after treatment).

If you are worried about your health at any time, contact your GP or medical team. Don't wait for your next appointment.

Relapsed and refractory hairy cell leukaemia

The first treatment for hairy cell leukaemia is usually very effective and people often stay in remission for many years. However, hairy cell leukaemia often comes back (**relapses**) eventually, and needs more treatment. Occasionally, hairy cell leukaemia doesn't respond well to your first treatment. This is called 'refractory' hairy cell leukaemia. It is usually treated the same way as relapsed hairy cell leukaemia.

The treatment you have depends on how long your remission lasted.

If you experience a relapse more than 2 years after your first treatment, you are likely to have the same treatment as you had before, usually combined with **rituximab**.

If you experience a relapse less than 2 years after your first treatment, your medical team might recommend:

- The other chemotherapy drug, usually combined with rituximab. For example, if you had pentostatin as your first treatment, you are likely to have cladribine (plus rituximab) if you experience a relapse.
- A different targeted drug as part of a **clinical trial**.

Occasionally, your medical team might recommend having an operation to remove your spleen (a **splenectomy**). This is usually considered only if your spleen is very enlarged and causing serious problems.

Relapsed hairy cell leukaemia is usually treated successfully and most people achieve a complete response. However, if you experience another relapse, there are still good treatment options. These include:

- Rituximab combined with a different chemotherapy drug (for example, fludarabine or bendamustine).

- Moxetumomab pasudotox, a type of treatment called an **antibody–toxin conjugate**. This is an antibody joined to a toxin (a natural substance that is poisonous to cells). The antibody sticks to a protein called CD22 on the surface of B cells and carries the toxin directly to them. At the time of writing, moxetumomab pasudotox is being assessed to decide whether it should be licensed and made available on the NHS for people with relapsed or refractory hairy cell leukaemia.

Research and targeted treatments

Standard treatments are usually very successful for hairy cell leukaemia. Clinical trials continue to test new treatments, most often for people with hairy cell leukaemia that is difficult-to-treat.

Scientists are testing many different **targeted treatments** in **clinical trials**, including some treatments that are already approved for other types of cancer. New treatments that are being tested in people with hairy cell leukaemia include:

- **ibrutinib**, which blocks signals that B cells send to help them divide or stay alive
- **venetoclax**, which switches off the survival signals that keep some cancer cells alive
- **trametinib**, which blocks a protein called MEK that makes cancer cells grow and divide.

Some of these might be available to you through a clinical trial. If you are interested in taking part in a clinical trial, ask your doctor if there is a trial that might be suitable for you. To find out more about clinical trials or search for a trial that might be suitable for you, visit **Lymphoma TrialsLink**.

Hairy cell leukaemia variant

Hairy cell leukaemia variant (HCL-V) is a type of blood cancer that is similar to hairy cell leukaemia, although it's actually a completely separate disease. Despite its name, it is classed as a form of **non-Hodgkin lymphoma**. It is very rare.

Like classical hairy cell leukaemia, the abnormal cells in HCL-V look hairy under a microscope. The main difference is that the abnormal cells in classical hairy cell leukaemia have a genetic change called a *BRAF* mutation, but the abnormal cells in HCL-V do not.

People with HCL-V often develop a swollen **spleen**. This can cause pain, discomfort, or a feeling of fullness in your tummy (abdomen). Unlike classical hairy cell leukaemia, HCL-V typically causes very high white blood cell counts. Low levels of white blood cells called monocytes (monocytopenia) are unusual.

HCL-V can be more difficult to treat than hairy cell leukaemia. The first treatment is usually **rituximab** combined with a chemotherapy drug (typically cladribine or pentostatin). Some people have surgery to remove their spleen (a **splenectomy**), if it is very enlarged. Other treatment options include **targeted drugs**.

References

The full list of references for this page is available on our website. Alternatively, email **publications@lymphoma-action.org.uk** or call 01296 619409 if you would like a copy.

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