

# Treating Hairy Cell Leukaemia- A Quick Guide



## Contents

This is a brief summary of Treating hairy cell leukaemia. You will find more detailed information on the website.

In this information there are sections on

- Staging hairy cell leukaemia
- Statistics and prognosis
- Types of treatment for hairy cell leukaemia
- Chemotherapy
- Biological therapy for hairy cell leukaemia
- Surgery
- Treatment if hairy cell leukaemia comes back
- Follow up for hairy cell leukaemia
- Hairy cell leukaemia research
- Questions for your doctor

## Staging hairy cell leukaemia

Stage means how far a cancer has grown or developed when it is diagnosed. Most types of cancer have a numbered stage, but stage is not used much for HCL. Your doctor works out your treatment for HCL based on any symptoms you have and your general health and fitness.

There is a staging system for hairy cell leukaemia (HCL), but specialists generally don't use it. We have included it here in

case you hear about it. The system uses two symptoms as a way of measuring disease. Doctors call these markers. The markers for HCL are

- Your level of anaemia (your red blood cell count)
- The size of your spleen

There are 3 stages in this system, depending on whether you have either of these symptoms and on how severe they are. If both are normal, or only slightly abnormal, it is stage 1. Moderate abnormalities are stage 2. A very enlarged spleen and severe anaemia is stage

## Statistics and prognosis of hairy cell leukaemia

Outlook means your chances of getting better. Your doctor may call this your prognosis. The outlook for hairy cell leukaemia depends on a number of different factors including how advanced the HCL is when you are diagnosed, and how well it responds to treatment. For detailed information, you will need to talk this through with your own specialist.

Hairy cell leukaemia is usually a condition that develops slowly and can be kept under control for many years with treatment. You



may hear these periods called remission. Remission is when the disease is not active – you don't have symptoms and it doesn't show up in your blood samples. It can be possible to achieve a second remission with more chemotherapy if the hairy cell leukaemia comes back (relapses).

On the website, we have more information about the likely outcome of hairy cell leukaemia. The statistics are intended as a general guide only. Statistics for HCL are harder to estimate than for other, less rare cancers.

### **Types of treatment for hairy cell leukaemia**

The treatment for HCL varies depending on how far your leukaemia has developed, your symptoms, your age, general health and level of fitness.

#### **When to start treatment**

If you have symptoms at diagnosis you will need to begin treatment. If you don't have any symptoms, you probably won't need to start treatment. Some people don't need treatment for years. Starting treatment when you have no symptoms is unlikely to help your HCL. If your blood count changes, or if you develop symptoms of HCL, you will begin treatment.

HCL is a disease of remission and relapse. Treatment can get the leukaemia into remission, which means that the leukaemia has disappeared completely or is under control. During this time, you don't need any treatment. With HCL, remission can last for years. If the disease becomes active again, it is called a relapse and you will start treatment again. Many people with HCL will have years of remission with further treatment.

### **The main treatments for hairy cell leukaemia (HCL)**

There are different treatments for HCL. You may need just one treatment, or a combination. The main treatment for hairy cell leukaemia is chemotherapy. You may also have a type of biological therapy called rituximab, surgery or interferon alpha.

### **Chemotherapy for hairy cell leukaemia**

Chemotherapy means treatment with drugs that kill cancer cells. The drugs travel through the body in the bloodstream. Chemotherapy is the most common treatment for HCL. The drugs you are most likely to have are cladribine or pentostatin.

#### **Cladribine**

Most people have cladribine as an injection just below the skin every day for 5 days in a row. You can have the injections as an outpatient. Some people have cladribine for a week at a time, as a continuous drip. This means that you will have to stay in hospital. Most people have just one course of treatment with cladribine.

#### **Pentostatin**

You have pentostatin as an outpatient every 2 weeks, for as long as your doctor thinks you need it. Generally, the treatment lasts between 3 and 6 months. Each treatment takes around half an hour.

#### **Side effects of chemotherapy**

People don't all have the same side effects with the same drug. Some people have very few side effects at all. Common side effects of these chemotherapy drugs include a fall in the number of blood cells, and feeling tired and run down. Some people get a rash or feel sick



## Biological therapy for hairy cell leukaemia (HCL)

Biological therapy is treatment with substances made naturally in the body or that change the way particular substances work in the body. Doctors use a type of biological therapy called rituximab to treat hairy cell leukaemia. Rarely they may use another biological therapy drug called interferon.

### Rituximab (Mabthera)

Chemotherapy is the main treatment for hairy cell leukaemia. But if chemotherapy doesn't control the leukaemia or if it comes back after chemotherapy treatment your doctor may suggest that you have rituximab with chemotherapy. Rituximab works by seeking out a protein that is found on normal and leukaemic white blood cells (lymphocytes). Once it has found the lymphocytes, it sticks to them. The immune system then targets the cells and kills them. Normal lymphocytes then grow to replace those that are destroyed.

The most common side effect of rituximab is a reaction to the drug when you first have it. It may also cause fever, chills and shivering, feeling sick, an itchy rash, headaches, low blood pressure and wheezing.

### Interferon

If you can't have chemotherapy or rituximab for hairy cell leukaemia your doctor may prescribe interferon. It is a natural substance that our bodies produce as part of our immune response. Interferon works by helping to stop the leukaemia cells growing. It may also encourage the immune system to attack the cancer. You have it as an injection just under the skin, usually 3 times a week for a year.

The main side effects are flu symptoms during the first week or two of treatment. Interferon can also make you feel sick, lose your appetite, or feel very tired

## Surgery for hairy cell leukaemia

Doctors don't use surgery to treat hairy cell leukaemia (HCL) very often. But some people with HCL need an operation to remove their spleen. The spleen is quite a large organ that filters the blood, destroys and recycles worn out red blood cells. In HCL, it sometimes swells up and causes problems. An operation to remove the spleen is called a splenectomy.

### Before your operation

Before your operation your surgeon will explain what will happen during the operation. A physiotherapist will come and explain the breathing and leg exercises you will need to do after your operation.

To help prevent infection in the future, you will need to have some vaccinations before, or just after, the operation. This is because the spleen is part of your immune system. Without a spleen you will be more at risk of infection.

### Recovering from your operation

When you wake up, you will probably have some tubes in place. You will have a drip to give you fluids and drainage tubes from your wound to help it heal. You can usually go home after a week or so, or sooner if you have had keyhole surgery.

If you do not have a spleen, you are at risk of serious infection. You will have to take antibiotics for the rest of your life to help avoid infection. You should carry a card saying you have no spleen, in case of medical emergencies. If you are planning to



travel abroad, you may need extra vaccinations

### **Treatment if hairy cell leukaemia comes back**

If your hairy cell leukaemia (HCL) comes back after earlier treatment, you will have what doctors call second line treatment. Most people have chemotherapy with either cladribine or pentostatin as a first treatment. The second line treatment you have will depend on the time since your last treatment, and what treatment you have already had.

If your last treatment was more than 3 to 5 years ago, you might have the same chemotherapy drug again. But if it was less than 3 years, you may have a different drug.

As well as the chemotherapy your doctor may suggest giving Rituximab. Rituximab is a type of biological therapy called a monoclonal antibody. It works by seeking out a protein that is found on some normal and leukaemic white blood cells, including hairy cell leukaemia cells. The protein is called CD20. Once it has found the CD20 positive cells, Rituximab sticks to them. This helps other immune system cells to find them and kill them.

If you are not well enough to have chemotherapy or rituximab your doctor may suggest that you have interferon. Interferon is a type of biological therapy that helps the immune system to destroy the leukaemia cells.

### **Follow up for hairy cell leukaemia**

Even after a long period where there is no sign of the leukaemia (remission) there is a possibility that it could return. Because of this your doctor will arrange for you to

have regular check ups. These will continue for some years, and possibly for life.

### **How often you will have check ups**

If you are not having any treatment, you will usually only need to see the doctor every 3 to 6 months. If you are having treatment, you will need to see your doctor more often. If you stay well, after a couple of years you may only need to go once a year.

### **What will happen during your check ups**

You will have blood tests and your doctor will want to know how you are feeling. You may have a physical examination as well. If you are worried, or notice any new symptoms between appointments, let your doctor or nurse know straight away. You don't have to wait until the next appointment.

### **Coping with worry**

Many people worry about going for their check ups. They can bring back all the worry about your cancer. You may find it helpful to tell someone close to you about how you are feeling.

It is quite common for people to have counselling after cancer treatment. To find out more about counselling, look in the coping with cancer section of the website.

### **Hairy cell leukaemia research**

All treatments must be fully researched before they can be adopted as standard treatment for everyone. This is so that we can be sure they work better than the treatments we already use. And so we know that they are safe.

Firstly, treatments are developed and tested in laboratories. Only after we know that they are likely to be safe to test are they



tested in people, in clinical trials. As hairy cell leukaemia is a rare cancer, it is more difficult to do trials. Talk to your doctor if you are interested in a new treatment and they will be able to tell you if it is appropriate for you.

There is research looking into diagnosing hairy cell leukaemia. There is also research into treating it with new chemotherapy drugs and biological therapies.

### Questions for your doctor about treating hairy cell leukaemia

- What sort of treatment do I need?
- Why would you recommend that treatment?
- What do you expect the treatment to do for me?
- Is there a choice of treatments?
- Will I be able to have another treatment if my disease flares up or returns?
- How long will my treatment last?
- What will the side effects be?
- Can you arrange for me to have a second opinion?
- Is there a clinical trial that I could join?
- Will I still be able to have children after treatment?
- How often will you see me when my treatment is finished?
- When will I be able to go abroad for a holiday?

### More information

For more information about hairy cell leukaemia, visit our website <http://cancerhelp.cancerresearchuk.org>

You will find a wide range of detailed, up to date information for people affected by cancer, including a clinical trials database that you can search for trials in the UK. You can view or print the information in a larger size if you need to.

For answers to your questions about cancer call our Cancer Information Nurses on **0808 800 4040**  
9am till 5pm Monday to Friday

Adapted from Cancer Research UK's Patient Information Website CancerHelp UK in April 2012. CancerHelp UK is not designed to provide medical advice or professional services and is intended to be for educational use only. The information provided through CancerHelp UK and our nurse team is not a substitute for professional care and should not be used for diagnosing or treating a health problem or disease. If you have, or suspect you may have, a health problem you should consult your doctor. © Cancer Research UK 2012. Cancer Research UK is a registered charity in England and Wales (1089464) and in Scotland (SC041666)