Hairy Cell Leukaemia (HCL)

A Guide for Patients

Leukaemia Care
YOUR Blood Cancer Charity

Introduction

Being diagnosed with hairy cell leukaemia (HCL) can be a shock, particularly when you may have never heard of it. If you have questions about HCL – what causes it, who gets it, how it affects your body, what symptoms to expect and likely treatments – this booklet covers the basics for you.

In this booklet, you'll also find useful advice about how to get the best from your haematologist, plus practical advice on how to help important people in your life understand such a rare condition. For more information, talk to your haematologist or clinical nurse specialist.

This booklet was originally written and subsequently revised by Ken Campbell MSc. It was then peer reviewed by Dr Manos Nikolousis & Dr Shankara Paneesha, Consultant Haematologists at the University Hospitals Birmingham NHS Foundation Trust and Nicola Shepherd, Clinical Nurse Specialist at Guy's Hospital. The review was conducted by Lisa Lovelidge and we are also grateful to Frank Mokrysz and Carina Patterson for their valuable contributions as patient reviewers. This booklet has since been updated by our Patient Information Writer, Isabelle Leach.

If you would like any information on the sources used for this booklet, please email **communications@leukaemiacare.org.uk** for a list of references.

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In this booklet

About Leukaemia Care

Leukaemia Care is a national charity dedicated to ensuring that people affected by blood cancer have access to the right information, advice and support.

Our services

Helpline

Our helpline is available 8:30am – 5:00pm Monday - Friday and 7:00pm – 10:00pm on Thursdays and Fridays. If you need someone to talk to, call **08088 010 444**.

Alternatively, you can send a message via WhatsApp on **07500068065** on weekdays 9:00am – 5:00pm.

Nurse service

We have two trained nurses on hand to answer your questions and offer advice and support, whether it be through emailing **nurse@leukaemiacare.org.uk** or over the phone on **08088 010 444**.

Patient Information Booklets

We have a number of patient information booklets like this available to anyone who has been affected by a blood cancer. A full list of titles – both disease specific and general information titles – can be found on our website at **www.leukaemiacare.org.uk/support-and-information/help-and-resources/information-booklets/**

Support Groups

Our nationwide support groups are a chance to meet and talk to other people who are going through a similar experience. For more information about a support group local to your area, go to **www.leukaemiacare.org.uk/support-and-information/support-for-you/find-a-support-group/**

Buddy Support

We offer one-to-one phone support with volunteers who have had blood cancer themselves or been affected by it in some

way. You can speak to someone who knows what you are going through. For more information on how to get a buddy call **08088 010 444** or email **support@leukaemiacare.org.uk**

Online Forum

Our online forum, **www.healthunlocked.com/leukaemia-care**, is a place for people to ask questions anonymously or to join in the discussion with other people in a similar situation.

Patient and carer conferences

Our nationwide conferences provide an opportunity to ask questions and listen to patient speakers and medical professionals who can provide valuable information and support.

Website

You can access up-to-date information on our website, **www.leukaemiacare.org.uk**.

Campaigning and Advocacy

Leukaemia Care is involved in campaigning for patient well-being, NHS funding and drug and treatment availability. If you would like an update on any of the work we are currently doing or want to know how to get involved, email **advocacy@leukaemiacare.org.uk**

Patient magazine

Our magazine includes inspirational patient and carer stories as well as informative articles by medical professionals: **www.leukaemiacare.org.uk/communication-preferences/**

What is hairy cell leukaemia?

Hairy cell leukaemia (HCL) is a rare type of leukaemia in which abnormal, mature B-lymphocytes (also known as B-cells) are present in the bone marrow, spleen and blood. B-cells are a type of white blood cell. The main characteristic of the B-cells in HCL is the hair-like projections on their surface which distinguish them from normal B-cells and those in other chronic B-cell leukaemias.

HCL develops slowly and usually responds well to treatment. Most people diagnosed with HCL are treated successfully and have a normal life expectancy.

Classical HCL

In classical HCL, the bone marrow creates too many defective hairy cell lymphocyte cells. Because these lymphocytes are abnormal, they cannot defend the body against infections. In addition, the bone marrow is unable to make enough normal blood cells as it is overcrowded with abnormal hairy cell lymphocytes. Hairy cells are found in the blood of almost 95% of cases.

HCL variants

HCL belongs to a varied group of B-cell disorders which include:

- The hairy cell variant (HCL-V)
- Splenic marginal zone lymphoma
- Splenic diffuse red pulp small B-cell lymphoma
- The VH4-34 molecular HCL variant

All these variants have hairy cells, but their clinical symptoms and genetic profiles are different.

HCL-V was thought to be a variant of HCL but is now considered by haematologists to be a separate disease which does not behave like HCL and is not treated in the same way. HCL-V is even rarer than classical HCL and can develop at any age, although it is more common in older people. If you have HCL-V, your haematologist will explain what this means and discuss your treatment with you.

In this booklet, we will be talking about classical HCL only.

Who is affected by HCL?

HCL is considered a rare disease as it affects less than 0.3 people in every 100,000 per year. It represents only 2% of adult leukaemias. There are about 1600 new cases of HCL diagnosed in Europe each year and the incidence of HCL is greater in Caucasians.

HCL is very rare in children and young adults, with a median age at the time of diagnosis of 52 years. It is more common in men than women, with a 4:1 ratio.

What causes HCL?

While the exact cause of HCL is difficult to determine, factors which are definitely known to increase the chance of developing the condition are older age and being male. You cannot catch HCL from someone who has it and you cannot pass HCL onto your children.

An important discovery has been that up to 95% of hairy cells have a specific mutation of a gene called B-raf proto-oncogene (BRAF). BRAF is a gene which helps to control cell division. Mutations of the BRAF gene have been reported in several types of cancer, including HCL.

The identified BRAF V600E mutation is absent from the lymphocytes of the other chronic B-cell leukaemias. This mutated gene cannot be passed on by parents; the gene is only abnormal in your hairy cells and not in your normal cells.

Symptoms of HCL

Some people do not have any symptoms when they are diagnosed with HCL and it is invariably identified following abnormal results during a routine blood test. Nevertheless, most people initially have minor symptoms which get gradually worse as the number of hairy cell lymphocytes increase in the bone marrow.

Classical HCL is characterised by:

- An enlarged spleen
- A decrease in all blood cell counts (red blood cells, white blood cells and platelets)
- Involvement of the bone marrow with fibrosis

When all the types of blood cells are lower than normal, this is called pancytopenia and it is present in the majority of patients with HCL. The decrease in the level of all the blood cells is due to the abnormal HCL cells occupying the bone marrow and taking the space of normal cells.

These low blood counts will cause most of the symptoms you might develop:

- Low levels of red blood cells (anaemia) can cause tiredness, fatigue and shortness of breath. This is present in approximately 70% of patients.
- Low levels of platelets will make bruising and bleeding more likely. This is present in about 80% of patients.
- Low levels of neutrophil white blood cells will make you more prone to frequent infections with fever. This is present in about 75% of patients.
- Low levels of monocyte white blood cells which is characteristic of HCL and is observed in 90% of patients with HCL.

An enlarged spleen is present in 70% to 90% of patients with HCL and can also cause the following:

- Abdominal (stomach) pain
- Swelling at the top-left of your abdomen
- Loss of appetite or feeling full after eating
- Increased urinating

In addition to the symptoms above, other symptoms which may be present in patients with HCL include:

- Excessive sweating, particularly at night
- Painless lumps in the neck, underarm, stomach or groin. These are lymph nodes which are part of the body's immune system. Lymph nodes contain lymphocytes which produce antibodies and macrophages to digest dead cells
- Weight loss

How is HCL diagnosed?

Development of blood cells

Each day more than a trillion new blood cells are produced in the bone marrow to replace those that have worn out. Approximately one in 5000 cells in the bone marrow is a blood-forming stem cell which can either divide to develop into one of the working blood cells or produce more stem cells. The production of new blood cells is very closely controlled so that it is balanced with the loss of worn-out cells or cells lost by bleeding or damage.

A blood stem cell in the bone marrow may become a myeloid cell or lymphoid cell.

A myeloid cell develops into one of three types of mature blood cells:

- Red blood cells that carry oxygen and other substances to all tissues of the body.
- Platelets that form blood clots to stop bleeding.
- White blood cells that fight infection and disease.

A lymphoid cell becomes one of three types of lymphocyte white blood cell:

- B-cells that make antibodies to help fight infection.
- T-cells that help the B-cells make the antibodies to fight infection.
- Natural killer cells (NK-cells) that attack cancer cells and viruses.

White blood cells can be granulocytes and include:

- Neutrophils that protect the body against bacterial infections and inflammation.
- Eosinophils that protect against parasites and allergens.
- Basophils that create the inflammatory reactions during an immune response.

Other white blood cells include:

- Lymphocytes that recognise bacteria, viruses and toxins, to which they produce antibodies.
- Monocytes that clear any infection products from the body.

Blood counts and films

If you do not have any symptoms when you are first diagnosed, your HCL will have been picked up following a routine full blood test which will have shown low levels of all your blood cells (red blood cells, white blood cells and platelets). Examination of your blood film, which is a thin layer of blood smeared on a glass microscope slide, generally shows hairy cells in the blood and a low level of monocyte white blood cells, which is typical of HCL. In addition, macrocytosis, which is an enlargement of the red blood cells, may be present.

Immunophenotyping

This process uses antibodies made by the patient to identify the types of antigens or markers on the surface of their leukaemia cells in blood or bone marrow samples. According to which antibodies are present on the leukaemia cells, it is possible to identify the type of leukaemia.

Immunophenotyping in patients with HCL can show antibodies for CD11c, CD25, CD103 and CD123, which are quite specific to HCL. In addition, antibodies for the markers CD19, CD20 or CD22 may be present. CD200 is a specific antibody marker which is highly present in HCL.

Bone marrow investigations

Examination of blood films and immunophenotyping can achieve a diagnosis of HCL in most cases. However, the diagnosis of HCL must be confirmed, as is the case of most leukaemias, with bone marrow investigations.

This is usually done to confirm the classical signs of HCL. During these investigations, you may have either a bone marrow aspiration or a bone marrow biopsy, or both. You are awake for the biopsy but you will have local anaesthetic to numb the area meaning that the procedure should not be overly painful. During a bone marrow aspiration, the doctor takes some bone marrow cells up into a needle and syringe, generally from your hip bone. A bone marrow biopsy uses a larger trephine needle to

How is HCL diagnosed? (cont.)

remove a one to two-centimetre core of bone marrow in one piece. The samples are then sent to the laboratory for testing. Both samples will be looked at later under a microscope.

Gene mutation analysis

Not only does the gene mutation analysis help in diagnosing difficult cases of HCL, but it will also inform your haematologist on your prognosis and how to treat you. You will be tested for the BRAF V600E mutation, the IGHV4-34 gene and your IGHV gene status:

- The BRAF V600E mutation has been reported in about 90% of patients with classical HCL, with 10% having the IGHV4-34 mutation or no mutation at all.
- The BRAF V600E mutation is not present in patients with HCL-V, other B-cell leukaemias or lymphomas.
- A mutated immunoglobulin heavy chain variable region (IGHV) gene is also present in approximately 90% of patients with HCL.
- The IGHV3-23 gene is the most common gene found in patients with HCL.

Therefore, the diagnosis of HCL is based on the combination of the symptoms experienced by the patient, microscopic examination of blood, lymph nodes or bone marrow, and the findings from the immunophenotyping and the gene mutation analysis.

Treating HCL

Overview of treatment

For patients who do not have any symptoms, the watch and wait approach is a good option because evidence has shown that early treatment does not offer any benefit in terms of survival for these patients. This option is also called active monitoring.

For patients with symptoms, antimetabolite cancer drugs have markedly improved the treatment of HCL. They work by interfering with DNA synthesis to prevent the growth or reproduction of cancer cells. The drugs cladribine and pentostatin (less commonly given) have achieved complete response rates in up to 90% of patients with HCL. Complete response is defined by blood counts returning to normal, disappearance of hairy cells from the blood and bone marrow, and a reduction of at least 50% of an enlarged spleen.

Patients have also had treatment-free periods of 10 years or more.

Watch and wait

If you do not have any symptoms when you are first diagnosed with HCL and your blood counts are not very low, your haematologist may suggest a watch and wait approach. This is generally the case for around 10% of people with HCL. This usually involves regular check-ups and blood counts, as well as your haematologist advising you on ways to live a healthy lifestyle.

If you would like more information on watch and wait, you can download our booklet from our website **www.leukaemiacare.org.uk**. Alternatively, you can order a hard copy by emailing Patient Services at **support@leukaemiacare.org.uk** or calling the helpline on **08088 010 444.**

If symptoms develop and the disease progresses, you may then be started on a suitable treatment.

Active treatment

Chemotherapy

Treating HCL (cont.)

Chemotherapy is the use of anticancer drugs to destroy the cancer cells. It has a very high success rate in the treatment of HCL. It does not cure the disease but it gives very good control and most patients can expect a normal or near-normal lifespan.

Purine analogues

The main drugs used to treat HCL are cladribine and pentostatin, which are antimetabolite purine analogues. Antimetabolites are drugs which interfere with the production of DNA, and therefore prevent the growth or reproduction of cancer cells. Antimetabolite cancer drugs can be divided according to their structure and function as:

- Purine analogues
- Folic acid analogues
- Pyrimidine analogues
- Cytidine analogues

The structure of these analogues are similar to the proteins used to make DNA but they differ enough to interfere with the process when they are incorporated into the DNA.

HCL responds well to both these drugs, although cladribine is the preferred choice because it can be given as a single five- to seven-day course, whereas pentostatin is given every other week for three to twelve months. Both drugs can be given into a vein (intravenously). Cladribine can also be given as injections just under the skin (subcutaneously).

Cladribine and pentostatin can affect the body's immune system making you more vulnerable to infections. If you need a blood transfusion, you will be given irradiated blood to reduce your chance of infections and complications. Irradiated blood is not radioactive and is just as safe and effective as any other blood transfusion. You may also be given antiviral drugs to prevent some viral infections.

For patients with HCL who do not respond to cladribine or pentostatin, other drugs are available:

- Another antimetabolite purine analogue that can be used for the treatment of HCL is fludarabine which is also used to treat some lymphomas, often

in combination with rituximab.

- Biological treatments such as interferon, which was widely used to treat HCL before cladribine and pentostatin were available.
- The monoclonal antibody rituximab can be added to cladribine or pentostatin to improve their efficacy.

The side effects of cladribine and pentostatin are similar. They include:

- Low levels of neutrophil white blood cells (potentially causing a fever)
- Nausea and vomiting
- Dizziness
- Headaches
- Fatigue
- Nerve damage (rare)

The aim of active treatments is to reduce the number of abnormal hairy cells to as low as possible. It is generally accepted that standard treatment will not cure HCL; but it does offer a very high chance of a normal, good quality of life.

Recommended first-line treatment for patients with HCL is cladribine or pentostatin and many people remain in remission for a long time without further treatment. However, approximately half of patients will relapse and require more treatment.

If the relapse is more than two years after the first treatment, re-treatment usually involves the same chemotherapy drug. If the relapse is less than two years after the first treatment, another chemotherapy drug is likely to be used. Additionally, rituximab (Mabthera) can be added to the chemotherapy with very good results. Other treatment options for a relapse are newer drugs or a bone marrow stem cell transplant.

Biological treatments

Biological treatments seek to make the cancer cells more recognisable to your immune system. Once your immune system identifies the cancer cells as intruders, it can set about destroying them.

Treating HCL (cont.)

There are two types of biological treatments that are used in the treatment of patients with HCL:

- Rituximab
- Interferon alpha

Rituximab

Rituximab is a monoclonal antibody which is widely used to treat chronic lymphocytic leukaemia (CLL) and lymphomas. Monoclonal antibodies are drugs that recognise, target and stick to particular proteins on the surface of cancer cells. They then stimulate the body's immune system to destroy these cells.

As a monoclonal antibody, rituximab binds to the CD20 protein on the surface of B-cells and destroys the HCL B-cells by targeting them specifically. Rituximab has limited action when given on its own, but is very effective when combined with cladribine and pentostatin.

In patients with HCL, cladribine followed by rituximab has achieved complete remission rates of up to 100%, showing a high efficacy of the combination of chemotherapy and immunotherapy. Five-year overall survival of 96.8% has been reported. The regimen is well-tolerated, with no severe or unexpected toxicity.

The most common side effects with rituximab are similar to those for cladribine and pentostatin. Patients often have rashes or other mild reactions following administration of their first dose of rituximab but this is usually easily controlled and is less common with subsequent doses.

Interferon Alpha

Interferon alpha is a substance which occurs naturally in the body and reduces the production of bone marrow cells. It can be made into a medicine to be given subcutaneously which reduces the rate at which blood cells are made, especially the abnormal hairy B-cells. Interferon is less used nowadays because newer chemotherapy drugs are more effective.

Interferon alpha is particularly useful for patients with HCL and fever due to an infection. Normally patients with an infection should

be treated before using purine analogues, but if the infection cannot be controlled, then interferon alpha can be used in the short term. Interferon alpha can be helpful for pregnant women who cannot take purine analogues.

Side effects with interferon alpha include:

- Flu-like symptoms
- Headaches
- Vision disturbances
- Depression
- Liver and thyroid disease

However, it does not increase the risk of leukaemia and can be used in pregnancy.

Splenectomy (surgical removal of the spleen)

Removal of the spleen was a standard treatment for HCL before purine analogues became available. It may be an option if your enlarged spleen is painful and your blood count results are not too low. Splenectomy can also be used for truly refractory HCL, although this is usually a last resort if all medications have failed. Refractory HCL is a type of HCL which does not respond to standard treatment. A splenectomy is also an option for pregnant women in order to postpone the administration of chemotherapy.

Despite the fact that removal of the spleen does not achieve remission, the blood counts return to normal in 40% to 70% of patients and five-year overall survival rates are 60% to 70%.

If you have had a splenectomy, it may take some time for it to improve your condition, so you will have a period of at least six months where you do not have any treatment to allow your haematologist to tell whether the splenectomy is controlling your HCL. After this time, you will have regular blood tests and will start drug treatment if these show that your HCL is progressing.

Treatment of relapsed HCL

Approximately 50% of patients relapse in the first five years after first-line treatment. Cladribine or pentostatin can still be effective

Treating HCL (cont.)

for the treatment of relapsed HCL. If the relapse is more than five years after the first treatment, treatment with cladribine or pentostatin is often used again. If the relapse is less than five years after the first treatment, another purine analogue such as fludarabine or a new drug such as a BRAF V600E inhibitor can be used.

New treatments and treatments on the horizon

Because standard treatment with purine analogues is so effective as a treatment for HCL, most studies of new treatments are concentrating on treatment of relapsed or refractory HCL. The most promising treatments include BRAF inhibitors and immunotoxins.

- **Vemurafenib** – A drug which targets the BRAF V600E mutation and has shown to be effective in patients who have experienced multiple relapses after purine analogue treatment or who do not respond to purine analogues.
- **Dabrafenib** - A BRAF inhibitor which is currently approved for the treatment of patients with BRAF V600E mutation-positive melanoma and non-small cell lung cancer.
- **Trametinib** – A mitogen-activated extracellular signal-regulated kinase (MEK) inhibitor which has a specific antigen for the BRAF V600E mutation.
- **Ibrutinib** – One of the first oral Bruton's tyrosine kinase inhibitors. It stops the enzyme working that is crucial for the survival of leukaemia cells in various B-cell cancers.
- **Immunotoxins** – This is where a bacterial toxin (poison generated by a bacteria) is fused to a monoclonal antibody. The monoclonal antibody targets and sticks to specific proteins on the surface of cancer cells such as CD22 which is present on nearly all hairy B-cells. Preliminary results in patients with HCL who have relapsed are showing promising results and no dose-related side effects have been seen.

More research and clinical trials

are needed before deciding the role of these drugs in the treatment for HCL.

What is the prognosis for HCL?

To date, treatment for patients with HCL has not provided a cure; however, it does provide the possibility of a normal life expectancy.

If you do not have any symptoms caused by an enlargement of your spleen and your blood cell counts are not greatly reduced, you may not require any treatment. If this is the case, you may be managed using the watch and wait approach with regular blood counts and physical check-ups.

Patients with symptoms of HCL should be treated with chemotherapy (such as purine analogues). The purine analogues, cladribine and pentostatin, can achieve complete responses in up to 70% to 90% of patients with HCL, with a survival rate of 10 years ranging between 85% and 100% of patients.

Your prognosis will be influenced by your individual situation and medical history, as well as the ways you respond to treatment. HCL affects people differently, and a patient's prognosis may vary depending on a number of factors such as:

- Genetic profile
- Blood counts
- Certain symptoms
- Age

A poorer prognosis is typically associated with the following:

- Absence of the BRAF V600E mutation
- Presence of the IGHV4-34 mutation or no mutation at all
- Unmutated IGHV gene (no mutation in the IGHV gene)
- Not responding to treatment with cladribine or pentostatin
- Enlarged spleen greater than 3cm on examination
- Enlarged lymph nodes
- Presence of CD38 antibody on hairy B-cells
- Changes in blood counts:

Treating HCL (cont.)

- Increased levels of white blood cells of more than $10x10^9/L$
- Hairy B-cells in the blood of more than $5x10^9/L$
- Haemoglobin below 10g/dL (normal range for men is 13.0g/dL to 18g/dL, and for women is 11.5g/dL to 16.5g/dL)
- Platelets less than $100x10^9/L$ (normal range is between $150x10^9/L$ and $450x10^9/L$)

HCL is a chronic condition, so it is important to see your haematologist regularly and to report any new or different symptoms. Your haematologist will tell you how often they need to see you as it will be based on your specific circumstances.

Patients with HCL can expect a favourable future. The challenge is to identify it as soon as possible and to treat it appropriately with purine analogues when required.

Living with HCL

After a diagnosis of HCL, you may find that it affects you both physically and emotionally. This chapter will talk about both of these aspects.

Emotional impact of HCL

Being told you have cancer can be very upsetting. Although the outlook for many HCL patients is a positive one, it is a rare type of leukaemia and, because of this, you may need emotional, as well as practical, support. Being diagnosed with a rare disease can affect the whole of you, not just your body, and can impact you emotionally at any point of your journey. It is likely that you will experience a range of complex thoughts and emotions, some of which may feel strange or unfamiliar to you. It is important to know that these feeling are all valid and a normal response to your illness.

It is important to remember that with current treatments, you can expect a good response and to live a long, normal life.

Looking after you

You can live a long and normal life with HCL but you may want to make changes to your lifestyle to try to stay as well as possible after your diagnosis and during treatment. Don't try to change too much at once. Adopting a healthy way of living is about making small, manageable changes to your lifestyle.

A healthy lifestyle includes having a well-balanced diet and being physically active. With some of the side effects you may be experiencing, the idea of getting out and being active may be the last thing you want to do, but it is important to try and stay as active as possible to make you feel better and help reduce some of your symptoms or side effects.

One of the most commonly reported side effects of the treatment of HCL is fatigue. This isn't normal tiredness and doesn't improve with sleep.

Some general tips on how to deal with fatigue include:

- Have a regular lifestyle – try

Living with HCL (cont.)

going to bed and waking up approximately the same time every day and try to avoid lying in.

- Take part in regular, gentle exercise to maintain your fitness levels as much as possible.
- Reserve your energy for what you find important and build rest periods around those times.
- Before going to bed, avoid stimulants such as alcohol, coffee, tea or chocolate, or using laptops, tablets or mobile phones.
- Keep your bedroom quiet and at a comfortable temperature.
- Talk about your worries with family, friends, or your doctor or clinical nurse specialist, or patient support groups.

Practical support

Work and finances

Being diagnosed with HCL can sometimes lead to difficulties relating to your work life. Your diagnosis may lead to temporary sick leave or a reduction in working hours, but it can also mean that you have to stop work altogether. You may need to make an arrangement with your employer for times when you may need to go into hospital or for those times when you may not be well enough to go into work.

Your consultant, GP or clinical nurse specialist can arrange letters to confirm your diagnosis and the effects it may have on your work life to your employer. It is often worth taking time to explain HCL to your employer, as it is likely they will never have heard of the disease.

It is important for you to know that people with any form of cancer are covered by law by the Equality Act. This means that legally your employer cannot discriminate against you and must take reasonable arrangements and adjustments relating to your disease.

If you would like advice about some of the financial help available to you, then you can speak to our Patient Advocacy team on **08088 010 444**. Alternatively, Macmillan has published a booklet about financial support following a

diagnosis of cancer that might be useful to you. They can also give you personal advice over the phone via their helpline at **0808 808 0000** and you can discuss which benefits you are eligible for. Some Macmillan centres can arrange face-to-face meetings with a benefits advisor. They can also provide financial assistance in the form of grants – ask your clinical nurse specialist in the hospital how to apply.

As HCL is a cancer, you will also be entitled to apply for a medical exemption certificate which means that you are entitled to free NHS prescriptions. Your GP or clinical nurse specialist at the hospital can provide you with the details of how to apply for this.

Talking about your HCL

Talking to your haematologist

HCL is a rare condition. It is important for you to develop a good working relationship with your haematologist so you are given the best treatment possible for you.

The following gives advice on working well with your haematologist:

- If it's an initial consultation, take along a list of your current medications and doses, and a list of any allergies you may have.
- If you have a complicated medical history, take a list of diagnoses, previous procedures and/or complications.
- Make a list of questions to take to your appointment. This will help the discussion with your haematologist.
- It can be useful to repeat back what you have heard so that you can be sure that you fully understood.
- Note information down to help you remember what was said.
- Be open when you discuss your symptoms and how you are coping. Good patient-doctor communication tends to improve outcomes for patients.

Other tips:

- Bring someone along to your appointment. They can provide support, ask questions and take notes.
- Don't be afraid to ask for a second opinion – most haematologists are happy for you to ask.

You need to tell your haematologist if...

You're having any medical treatment or taking any products such as prescribed medicines, over the counter treatments or vitamins. It is important to understand that treatments, including complementary therapies, which are perfectly safe for most people, may not be safe if you are being treated for HCL. Remember, if you choose to start any form of complementary therapy outside of your medical treatment, consult your haematology consultant or clinical nurse

specialist prior to beginning it. It is important to understand the difference between complementary therapies, used alongside standard treatment, and alternative therapies, used instead of standard treatment. There is no evidence that any form of alternative therapy can treat HCL.

Talking to other people

Telling people you have a rare condition like HCL can be hard to explain. You might find it useful to let your close family and friends, as well as your employer, know about your health condition. It might be easier to provide people with basic information leaflets about HCL (or a booklet like this one) if they want to know more in-depth details.

It is probably best to focus conversations on the symptoms you are experiencing, how the condition affects you and how you feel about it. Often people misunderstand and, unfortunately, it will mostly fall to you to educate them as best as you can. Where possible, it's advisable to let people know what you find helpful and unhelpful, in terms of what others say and do.

Often people make assumptions and do what they think helps. For example, saying you look well, recounting stories of others they know with a similar diagnosis or encouraging you to look ahead and stay positive isn't always what people really want to hear. In many ways, the more you communicate with them the better.

These points may help you:

- Explain that you have a condition that means your bone marrow does not function properly, and this affects the number of blood cells it produces.
- Explain your symptoms (maybe you are tired, or have a lot of pain).
- Explain what you need (maybe more help day-to-day, or someone to talk to).

You could also consider the following when telling people about your diagnosis:

- **Find out more** – Try to find out as much as you can about your condition, from reliable internet sources, charitable organisations or your

Talking about your HCL (cont.)

consultant haematologist and clinical nurse specialist. The more you know, the more you can share.

- **Have a print-out to hand** – It may help to have a factsheet or booklet to hand to share with family and friends. This will take the pressure off you having to remember everything they may want to know.
- **Explain your needs** – Try and be clear about what your needs may be. Perhaps you need help with the weekly food shop, help with cooking dinner or someone to drive you to and from appointments. You may find that friends and family are pleased that they can do something to help you.
- **Be open about how you feel** – Don't be afraid of opening up about how you feel, as people who care will want to help you as best they can. Talk as and when you feel comfortable, so those around you will know when you need them most.

Glossary

Anaemia

A medical condition in which the red blood cell count or haemoglobin level is less than normal.

Antibodies

A blood protein produced in response to and counteracting a specific antigen. Antibodies combine with substances which they recognise as alien, such as bacteria, viruses or foreign substances in the blood.

Antigen

A toxin or other foreign substance which induces an immune response in the body, especially the production of antibodies.

Blood Count

A common test to check a person's general health or to screen for anaemia.

B-lymphocyte

A type of lymphocyte white blood cell which produces antibodies to fight infection.

Bone Marrow

A soft blood-forming tissue that fills the cavities of bones and contains fat, immature and mature blood cells, including white blood cells, red blood cells and platelets.

Chemotherapy

A form of cancer treatment that uses one or more anticancer drugs as part of a standardised chemotherapy regime.

Chromosomes

Thread-like structures which carry the genes, and are located in the nuclei of every cell in the body. There are 46 chromosomes (23 pairs) in humans.

Chronic Leukaemia

A type of blood cancer that mainly affects the white blood cells. This tends to progress over many years.

Clinical Trial

A medical research study involving patients with the aim of improving treatments and their side effects. You will always be informed if your treatment is part of a trial.

Glossary (cont.)

Complete Remission

This means that tests, physical examinations and scans show that all signs of cancer are gone. There is no evidence of the disease.

Fatigue

Extreme tiredness and weakness rendering you unable to work or perform usual activities. Fatigue can be acute and come on suddenly, or chronic and persist over a longer period of time.

Full Blood Count (FBC)

A blood test that counts the number of different blood cells.

Genes

Genes are made up of DNA which stores the genetic information required to make human proteins.

Granulocyte

A type of white blood cell which is characterised by the presence of granules in the cytoplasm. They are important for fighting infection, particularly bacterial infections.

Hairy Cell Leukaemia

Hairy cell leukaemia is one of the rarest types of leukaemia, which is cancer of the B-lymphocyte white blood cells. It gets its name from the fine, hair-like strands around the outside of the cancerous cells, which are visible under a microscope.

Immunophenotyping

Immunophenotyping is used to help diagnose and classify the blood cell cancers, leukaemias and lymphomas, and to help guide their treatment. Antibodies are used to identify cells by detecting specific antigens on the surface of these cells, which are known as markers.

Leukaemia

A cancer of the bone marrow/blood with many different subtypes. Some forms are acute (develop quickly) and others are chronic (develop slowly). Leukaemia is an excess number of abnormal cells in the bone marrow, usually white blood cells, which stop the bone marrow working properly.

Lymphocyte

A type of white blood cell which forms part of the body's immune system.

Monoclonal Antibodies

Antibodies that are made by identical immune cells that are all clones of a unique parent cell. They therefore all bind to the same part of an antigen that is recognised by the antibody.

Mutation

The changing of the structure of a gene, resulting in a variant form which may be transmitted to subsequent generations. Mutations are caused by the alteration of single base units in DNA, or the deletion, insertion, or rearrangement of larger sections of genes or chromosomes.

Prognosis

An indication of how well a patient is expected to respond to treatment based on their individual characteristics at the time of diagnosis or other timepoint of the disease.

Refractory Disease

Refractory describes a disease or condition which does not respond to treatment.

Relapse

A relapse occurs when a patient initially responds to treatment, but after six months or more, the disease recurs.

Remission

A period of time when the illness is less severe or is not affecting someone because the cancer cells have been substantially decreased by treatment.

Spleen

The largest organ in the lymphatic system. Similar in structure to a large lymph node, it acts primarily as a blood filter and defends the body against infection.

White Blood Cells

White blood cells are one of the types of cells found in the blood and bone marrow, along with red blood cells and platelets. White blood cells create an immune response against both infectious

Glossary (cont.)

disease and foreign invaders. Granulocyte white blood cells, include the neutrophils (protect against bacterial infections and inflammation), eosinophils (protect against parasites and allergens) and basophils (create the inflammatory reactions during an immune response). Other white blood cells include the lymphocytes (recognise bacteria, viruses and toxins, to which they produce antibodies) and monocytes (clear infection products from the body).

Useful contacts and further support

There are a number of helpful sources to support you during your diagnosis, treatment and beyond, including:

- Your haematologist and healthcare team
- Your family and friends
- Your psychologist (ask your haematologist or CNS for a referral)
- Reliable online sources, such as Leukaemia Care
- Charitable organisations

There are a number of organisations, including ourselves, who provide expert advice and information.

Leukaemia Care

We are a charity dedicated to supporting anyone affected by the diagnosis of any blood cancer. We provide emotional support through a range of support services including a helpline, patient and carer conferences, support group, informative website, one-to-one buddy service and high-quality patient information. We also have a nurse on our help line for any medical queries relating to your diagnosis.

Helpline: **08088 010 444**
www.leukaemiacare.org.uk
support@leukaemiacare.org.uk

Blood Cancer UK

Blood Cancer UK is the leading charity into the research of blood cancers. They offer support to patients, their family and friends through patient services.

0808 2080 888
www.bloodcancer.org.uk

Cancer Research UK

Cancer Research UK is a leading charity dedicated to cancer research.

0808 800 4040
www.cancerresearchuk.org

Macmillan

Macmillan provides free practical, medical and financial support for people facing cancer.

0808 808 0000
www.macmillan.org.uk

Maggie's Centres

Maggie's offers free practical, emotional and social support to people with cancer and their families and friends.

0300 123 1801
www.maggiescentres.org

Citizens Advice Bureau (CAB)

Offers advice on benefits and financial assistance.

08444 111 444
www.adviceguide.org.uk

Leukaemia Care is a national charity dedicated to providing information, advice and support to anyone affected by a blood cancer.

Around 34,000 new cases of blood cancer are diagnosed in the UK each year. We are here to support you, whether you're a patient, carer or family member.

Want to talk?

Helpline: **08088 010 444**
(free from landlines and all major mobile networks)

Office Line: **01905 755977**

www.leukaemiacare.org.uk

support@leukaemiacare.org.uk

Leukaemia Care,
One Birch Court,
Blackpole East,
Worcester,
WR3 8SG

Leukaemia Care is registered as a charity in England and Wales (no.1183890) and Scotland (no. SC049802).
Company number: 11911752 (England and Wales).
Registered office address: One Birch Court, Blackpole East, Worcester, WR3 8SG

Leukaemia Care
YOUR Blood Cancer Charity