

Chronic Lymphocytic Leukaemia (CLL)

A guide for patients, families & whānau



our mission is to care, our vision is to cure



Leukaemia and Blood Cancer New Zealand is grateful to Roche Products (New Zealand) Limited for providing an unrestricted educational grant enabling the production of this booklet

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Introduction

This booklet has been written to help you and your family or whānau understand more about chronic lymphocytic leukaemia (CLL).

It is important to acknowledge that for many people CLL may never become a problem and they can continue to lead a normal life despite their diagnosis. For others, particularly where there is evidence of disease progression, CLL may represent a more serious condition.

If you or someone you care for has been diagnosed with CLL, you may be feeling anxious or a little overwhelmed. This is normal. Perhaps you have already started treatment or you are discussing different treatment options with your doctor and your family. Whatever point you are at, we hope that the information contained in this booklet is useful in answering some of your questions. It may raise other questions, which you should discuss with your doctor or specialist nurse.

You may not feel like reading this booklet from cover to cover. It might be more useful to look at the list of contents and read the parts that you think will be of most use at a particular point in time. We have used some medical words and terms that you may not be familiar with. Their meaning is either explained in the text, in the glossary of terms at the back of this booklet, or in the 'Dictionary of Terms' booklet.

Some people may require more information than is contained in this booklet. We have included some internet addresses that you might find useful. In addition, many of you will receive written information from the doctors and nurses at your treatment centre.

It is not the intention of this booklet to recommend any particular form of treatment to you. You need to discuss your circumstances at all times with your doctor and treatment team.

We hope that you find this booklet useful. There is a feedback form in the back of this booklet, please feel free to fill this in and return it to us to assist in the production of future editions.

Acknowledgements

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Leukaemia & Blood Cancer New Zealand also gratefully acknowledges Dr David Simpson (North Shore Hospital) and Dr Andrew Butler (Christchurch Hospital) for their assistance with the development of this booklet.

Leukaemia & Blood Cancer New Zealand

Leukaemia & Blood Cancer New Zealand (LBC) is the only organisation in New Zealand dedicated to supporting patients and their families living with leukaemia, lymphoma, myeloma and related blood conditions.

Since 1977, our work has been made possible through our fundraising events and the generous support we receive from individuals, companies, trusts and grants. We do not receive government funding.

LBC manages the New Zealand Bone Marrow Donor Registry, which works towards finding matched volunteer donors from New Zealand or overseas for New Zealand patients who need a bone marrow or stem cell transplant and who do not have a family donor. The registry maintains information on New Zealand donors and has access to a worldwide database of over 18 million donors.

Patient Support

Leukaemia & Blood Cancer New Zealand's Support Services provide personalised support programmes for patients and their families. This can include regular visits, phone or email contact, as well as face to face education and support programmes and an online information forum. We also provide a toll free number for advice, empathy and support.

Research

Research plays a critical role in building a greater understanding of blood cancers and conditions. LBC supports and funds investigation into these conditions. Improved treatments for patients can lead to increased survival rates.

Information

We provide vital information to patients, families, health professionals and the community to improve understanding about blood cancers and conditions.



Awareness

We work to increase public knowledge of blood cancers and conditions. This is achieved through specifically focused campaigns for the public, health professionals and health agencies.

Advocacy

We represent the needs of patients and their families to the government, related agencies and other relevant organisations.

Contacting us

Leukaemia & Blood Cancer New Zealand provides services and support throughout New Zealand. Every person's experience of living with a blood cancer or condition is different. Living with leukaemia, lymphoma, myeloma or a related blood condition is not easy, but you don't have to do it alone.



Call **0800 15 10 15** to speak to a local Support Services Coordinator or to find out more about the services offered by Leukaemia & Blood Cancer New Zealand. Alternatively, contact us via email by sending a message to info@leukaemia.org.nz or by visiting www.leukaemia.org.nz.

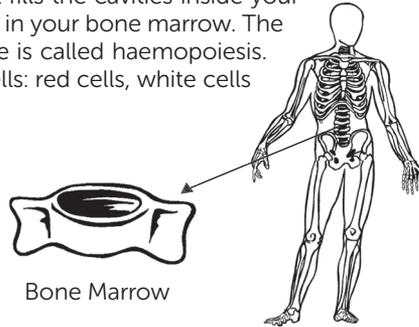
We welcome visitors to our offices in Auckland, Wellington and Christchurch. Please phone for an appointment.

Bone marrow, stem cells & blood cell formation

Bone marrow

Bone marrow is the spongy tissue that fills the cavities inside your bones. All of your blood cells are made in your bone marrow. The process by which blood cells are made is called haemopoiesis. There are three main types of blood cells: red cells, white cells and platelets.

As an infant, haemopoiesis takes place at the centre of all bones. As an adult, fewer new cells are needed – the marrow space in the arms and legs is replaced by fat, and active marrow is limited to the hips, ribs and breastbone (sternum).

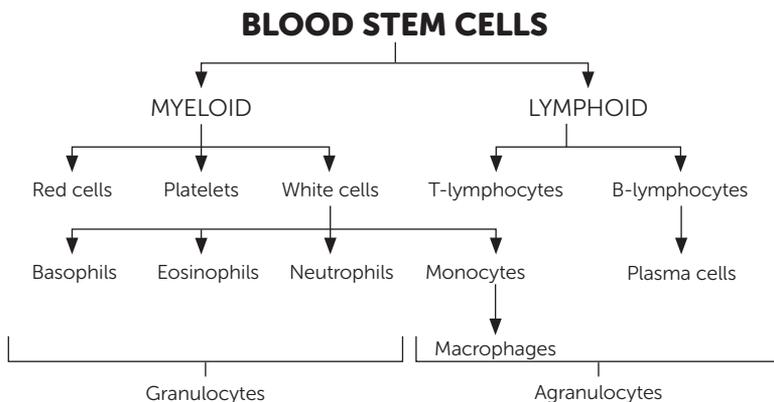


Bone Marrow

You might like to think of the bone marrow as the blood cell factory. The main workers at the factory are the blood stem cells. They are relatively few in number but are able, when stimulated, not only to replicate themselves, but also to grow and divide into slightly more mature stem cells called myeloid stem cells and lymphoid stem cells. These can multiply and mature further to produce all the circulating blood cells.

Myeloid ('my-loid') stem cells develop into red cells, white cells (neutrophils, eosinophils, basophils and monocytes) and platelets.

Lymphoid ('lim-foid') stem cells develop into two other types of white cells called T-lymphocytes and B-lymphocytes.



Blood

Blood consists of blood cells and plasma. Plasma is the straw coloured fluid part of the blood, which blood cells use to travel around your body.



Blood cells

Red cells and haemoglobin

Red cells contain haemoglobin (Hb), which transports oxygen from the lungs to all parts of the body. Haemoglobin also carries carbon dioxide to the lungs where it can be breathed out.

The normal haemoglobin range for a man is between 130 - 170 g/L

The normal haemoglobin range for a woman is between 120 - 160 g/L

Red cells are by far the most numerous blood cell and the proportion of the blood that is occupied by red cells is called the haematocrit. A low haematocrit suggests that the number of red cells in the blood is lower than normal.

The normal range of the haematocrit for a man is between 40 - 52%

The normal range of the haematocrit for a woman is between 36 - 46%

Anaemia is a condition caused by a reduction in the number of red cells, which in turn results in a low haemoglobin. Measuring either the haematocrit or the haemoglobin will provide information regarding the degree of anaemia.

If you are anaemic you will feel run down and weak. You may be pale and short of breath or you may tire easily because your body is not getting enough oxygen. In this situation a red cell transfusion may be given to restore the red cell numbers and therefore the haemoglobin to normal levels.

White cells

White cells, also known as leucocytes, fight infection. There are different types of white cells which fight infection together and in different ways.

Granulocytes:

Neutrophils	kill bacteria and fungi
Eosinophils	kill parasites
Basophils	work with neutrophils to fight infection

Agranulocytes:

T-lymphocytes	kill viruses, parasites and cancer cells; produce cytokines
B-lymphocytes	make antibodies which target microorganisms
Monocytes	work with neutrophils and lymphocytes to fight infection; they also help with antibody production and act as scavengers to remove dead tissue. These cells are known as monocytes when they are found in the blood and macrophages when they migrate into body tissues to help fight infection

If your white cell count drops below normal you are at risk of infection.

The normal adult white cell range is between $4.0 - 11.0 \times 10^9/L$

Neutropenia is the term given to describe a lower than normal neutrophil count. If you have a neutrophil count of less than $1.0 (1.0 \times 10^9/L)$ you are considered to be neutropenic and at risk of developing frequent and sometimes severe infections.

The normal adult neutrophil range is between $2.0 - 7.5 \times 10^9/L$

Platelets

Platelets are disc-shaped cellular fragments that circulate in the blood and play an important role in clot formation. They help to prevent bleeding. If a blood vessel is damaged (for example by a cut), the platelets gather at the site of the injury, stick together and form a plug to help stop the bleeding.

The normal adult platelet range is between $150 - 400 \times 10^9/L$

Thrombocytopenia is the term used to describe a low platelet count. If your platelet count is low, you are at higher risk of bleeding, and tend to bruise easily. Platelet transfusions are sometimes given to bring the platelet count back to a higher level. In certain situations, especially when patients are receiving some chemotherapy treatments, platelets may be transfused if the blood level falls below $10 \times 10^9/L$.

The normal blood counts provided here may differ slightly from the ones used at your treatment centre. You can ask for a copy of your blood results, which should include the normal values for each blood type.



Growth factors and cytokines

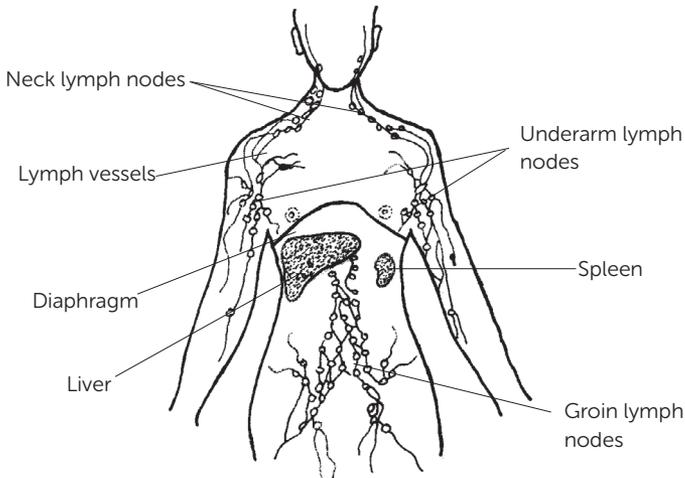
All normal blood cells have a limited survival in the circulation and need to be replaced on a continual basis. This means that the bone marrow remains a very active tissue throughout your life. Natural chemicals in your blood called growth factors or cytokines control the process of blood cell formation. Different growth factors stimulate the blood stem cells in the bone marrow to produce different types of blood cells.

Many growth factors can be made in the laboratory (synthesised) and are available for use in people with blood disorders. For example, granulocyte-colony stimulating factor (G-CSF) stimulates the production of white cells called neutrophils, while erythropoietin (EPO) stimulates the production of red cells. Unfortunately, drugs to stimulate platelet production have been less successful, but research is continuing in this area.

The lymphatic system

The lymphatic system is made up of a vast network of vessels, similar to blood vessels, that branch out into all the tissues of the body. These vessels contain lymph, a colourless watery fluid that carries lymphocytes, specialised white blood cells that fight infection. There are two types of lymphocytes, B-lymphocytes and T-lymphocytes (called B-cells and T-cells). These cells protect us by making antibodies and destroying harmful microorganisms like bacteria and viruses. As such, the lymphatic system forms part of the immune system, which protects our bodies against disease and infection.

Clusters of small bean-shaped organs called lymph nodes (also known as lymph glands) are found at various points throughout the lymphatic system. The lymph nodes, which are filled with lymphocytes, act as important filtering stations, cleaning the lymph fluid as it passes through them. Here bacteria, viruses and other harmful substances are removed and destroyed. When you have an infection, for example a sore throat, you may notice that the lymph nodes under your jawbone become swollen and tender. This is because the lymphocytes become activated and multiply in response to the virus or bacteria causing the infection.



The spleen (an organ on the left side of the abdomen), thymus (a gland found behind the breast bone), tonsils and adenoids (glands in the throat) and bone marrow (spongy material inside bones) all contain lymphatic tissue and are therefore considered to be part of the lymphatic system. Lymphatic tissue is also found in other parts of the body.

What is leukaemia?

Leukaemia is the general name given to a group of cancers that develop in the bone marrow. Under normal conditions the bone marrow contains a small number of healthy immature blood cells, sometimes called blast cells. These immature blood cells mature and develop into red cells, white cells and platelets, which are eventually released into the blood stream. Leukaemia originates in developing blood cells, which have undergone a malignant (cancerous) change. Instead of maturing properly these cells grow and multiply in an uncontrolled fashion and interfere with normal blood cell production in the bone marrow. Most cases of leukaemia originate in developing white cells. In a small number of cases leukaemia develops in other blood-forming cells, for example in developing red cells or developing platelets.

Types of leukaemia

There are several different types and subtypes of leukaemia.

Acute/chronic

Leukaemia can be either acute or chronic. The terms 'acute' and 'chronic' refer to how quickly the disease develops and progresses.

Acute leukaemia develops and progresses quickly and therefore needs to be treated as soon as it is diagnosed. It affects very immature blood cells, preventing them from maturing properly.

In chronic leukaemia there is an accumulation of more mature but abnormal white cells. It can occur at any age, but is more common in older adults and is rarely seen in children.

Myeloid/lymphoid

Leukaemia can also be either myeloid or lymphoid. The terms myeloid and lymphoid refer to the types of cell lineage in which the leukaemia first started.

When leukaemia starts somewhere in the myeloid cell line, it is called myeloid (myelocytic, myelogenous or granulocytic) leukaemia.

When leukaemia starts somewhere in the lymphoid cell line it is called lymphoblastic, lymphocytic, or lymphatic leukaemia. (See diagram of stem cell lines on page 5).

Therefore, there are four main types of leukaemia:

1. Acute myeloid leukaemia (AML)
2. Acute lymphoblastic leukaemia (ALL)
3. Chronic myeloid leukaemia (CML)
4. Chronic lymphocytic leukaemia (CLL)

Both adults and children can develop leukaemia but certain types are more common in different age groups.

There are separate booklets about the different types of leukaemia available from Leukaemia & Blood Cancer New Zealand.

What is chronic lymphocytic leukaemia (CLL)?

Chronic lymphocytic leukaemia (CLL) is a type of slow-growing leukaemia that affects developing B-lymphocytes. B-lymphocytes (also known as B-cells), are specialised white blood cells. Under normal conditions they produce immunoglobulins (also called antibodies) that help protect our bodies against infection and disease. In people with CLL, lymphocytes undergo a malignant (cancerous) change and become leukaemic cells.

It is important to emphasise that for many people with CLL, the condition can remain stable for many months or even years, and may have little if any impact on lifestyle or general health.

However, in other people, the leukaemic cells multiply in an uncontrolled way, live longer than they are supposed to and accumulate in the bone marrow, blood stream, lymph nodes, spleen and other parts of the body. These cells are abnormal and are unable to function properly. Over time, an excess number of lymphocytes crowd the bone marrow, and interfere with normal blood cell production. The bone marrow then produces inadequate numbers of red cells, normal white cells and platelets, making some people with CLL more susceptible to anaemia, recurrent infections and to bruising and bleeding easily. Circulating red cells and platelets can also be damaged by abnormal proteins made by the leukaemic cells.

CLL usually develops and progresses slowly, over many months or years. Most people have no symptoms of their disease when they are first diagnosed. In these cases, people often require no treatment for a long time, apart from regular check-ups with their doctor to carefully monitor their health. Others may need to be treated soon after they are diagnosed.

How common is CLL and who gets it?

Each year in New Zealand around 120 people are diagnosed with CLL. While CLL is a relatively uncommon type of cancer, it is the most common type of leukaemia diagnosed in New Zealand, and in the western world. The majority of people diagnosed with CLL are over the age of 60 (almost 80%). CLL is rare under the age of 40 and it has not been reported in children or adolescents. CLL is more common in men than women.

What causes CLL?

Many people who are diagnosed with CLL ask the question “why me?” Naturally, they want to know what has happened or what they might have done to cause their disease. In most cases the cause of CLL remains unknown. We do know that it is not contagious, that is, you cannot ‘catch’ CLL by being in contact with someone who has it.

Like other types of leukaemia, CLL is thought to arise from an acquired mutation (or change) in one or more of the special proteins, called genes that normally control the growth and development of blood cells. This change (or changes) will result in abnormal growth. The original mutation is preserved when the affected stem cell divides and produces a 'clone'; that is a group of identical cells all with the same defect.

Why gene mutations occur in the first place remains largely unknown. There are likely to be a number of, as yet, unidentified factors involved. In rare cases, CLL may result from an inherited defect in a gene, passed down from one generation to the next.

What are the symptoms of CLL?

As we mentioned earlier, many people have no symptoms when they are first diagnosed. In these cases the disease may be picked up by accident, for example during a routine blood test or physical examination. Other people may go to see their general practitioner (GP) because they have some troubling symptoms of their disease. These may include the following:

Symptoms caused by a lack of normal white cells and normal antibodies:

- Frequent or repeated infections

Symptoms of anaemia, due to a lack of normal red cells:

- Persistent tiredness and fatigue
- Weakness
- Shortness of breath with minimal exercise
- Looking pale

Symptoms caused by a lack of normal platelets:

- Bleeding or bruising more easily for no apparent reason
- Frequent or severe nose bleeds or bleeding gums
- The appearance of red or purple flat pinhead sized purple spots on the skin, especially on the legs initially. These are due to small superficial capillary bleeds known as petechiae ('pe-tee-kee-eye') and may have greater clinical importance than bruises.

CLL can cause a painless swelling of the lymph nodes (glands) in your neck, under your arms or in your groin. This is usually a result of lymphocytes accumulating in these tissues. Symptoms of an enlarged spleen (splenomegaly) are common and include feelings of discomfort, pain or fullness in the upper left-side of the abdomen. An enlarged spleen may also cause pressure on the stomach causing a feeling of fullness, indigestion and a loss of appetite. In some cases the liver may also be enlarged. Other symptoms of CLL may include excessive sweating, especially at night, fevers and unintentional weight loss.

Some of the symptoms described above may also be seen in other illnesses, including viral infections and most people with these symptoms don't have leukaemia. However, it is important to see your doctor if you have any unusual symptoms, or symptoms that don't go away so that you can be examined and treated properly.

Which doctor?

If your GP suspects that you might have leukaemia that requires treatment, you will be referred on to another specialist doctor called a haematologist for further tests and treatment. A haematologist specialises in the care of people with diseases of the blood, bone marrow and immune system.

For people with CLL not needing treatment, it is common to remain under the care of your GP, with regular blood tests as required.



How is CLL diagnosed?

CLL is diagnosed by examining samples of your blood in the laboratory. When you see your doctor about any of the symptoms above, the first thing he or she will probably do is take your full medical history, asking questions about your general health and any illness or surgery you have had in the past and give you a full physical examination. The doctor will look and feel for any swelling of the nodes in your neck, armpits and groin. Your abdomen and chest will also be examined for any signs of enlarged organs or fluid collection. The doctor will ask you about any other symptoms you might have and take some blood samples to check how well your bone marrow, liver and kidneys are functioning.

Full blood count

The first step in diagnosing CLL requires a simple blood test called a full blood count (FBC) or complete blood count (CBC). This involves taking a sample of your blood, usually from a vein in your arm, and sending it to the laboratory for examination under the microscope. The number of red cells, white cells and platelets, and their size and shape, is noted as these can all be abnormal in CLL.

In CLL, the lymphocyte count is abnormally high, and needs to be at least $5 \times 10^9/L$ for a diagnosis of CLL. Anaemia and thrombocytopenia (a lower than normal platelet count) are common in more advanced disease.

Your full blood count will be checked regularly both during and after treatment to see how well your disease is responding.

Immunophenotyping ('im-u-no-feen-o-typing')

Immunophenotyping, or flow cytometry tests, are commonly used to confirm a suspected diagnosis of CLL, and to distinguish it from other similar diseases. This technology uses the special markers, called antigens, found on the surface of cells. These antigens act like flags identifying the abnormal lymphocytes that are characteristic of CLL.

Antigens are commonly referred to as 'clusters of differentiation' or CD antigens followed by a number. In CLL certain B-cell antigens like CD5, CD19, CD20 and CD23 and other surface markers are almost always expressed on the leukaemic cells. The presence of these markers helps to define the exact type of CLL you have and distinguish it from other diseases that can resemble CLL. These include such diseases as prolymphocytic leukaemia, hairy cell leukaemia, mantle cell lymphoma and other types of lymphoma (cancer of the lymphatic system).

Cytogenetic ('cy-to-gen-etic') tests

Chromosomes are the organised structures of DNA inside our cells. A single piece of DNA contains many genes, which are our body's blueprint for life. Leukaemic cells have developed mutations in their genetic make-up which can be detected using cytogenetic testing. The two main types of tests are chromosome analysis, which examines chromosomes under a microscope, and FISH (fluorescent in situ hybridisation) which 'paints' the genes of interest with fluorescent dye.

Certain cytogenetic mutations, such as missing, extra or abnormal chromosomes help to confirm the subtype of CLL you have, the likely prognosis of your disease and the best way to treat it. Note: these mutations develop as your cells age, so this type of chromosomal change is only found in the leukaemia cells and is not inherited, that is passed down from parent to child.

Immunoglobulin (antibody) levels

Blood samples may also be taken to measure the levels of antibodies in your blood. People with low levels of normal antibodies may be more susceptible to repeated bacterial and viral infection and some may benefit from monthly intravenous immunoglobulin treatment to reduce the frequency of infections. Blood tests may be repeated at regular intervals to monitor your disease or see how well it is responding to treatment.

Bone marrow examination

A bone marrow examination (biopsy) is used in some cases to help confirm the diagnosis of CLL. It can also provide useful information about the likely course of the disease and to assess how well it is responding to treatment. It involves taking a sample of bone marrow, usually from the back of the iliac crest (hip bone) and sending it to the laboratory for examination under the microscope.

The bone marrow examination may be done in the hospital or outpatient clinic under local anaesthesia or, in selected cases, under sedation. A mild pain-killer is given beforehand and the skin is numbed using a local anaesthetic; this is given as an injection under the skin. The injection takes a minute or two, and you should feel only a mild stinging sensation.

After allowing time for the local anaesthetic to work, a long thin needle is inserted through the skin and outer layer of bone into the bone marrow cavity. A syringe is attached to the end of the needle and a small sample of bone marrow fluid is drawn out - this is called a 'bone marrow aspirate'. Then a slightly larger needle is used to obtain a small core of bone marrow which will provide more detailed information about the structure of the bone marrow and bone - this is known as a 'bone marrow trephine'.

If a sedative is used you might feel a bit drowsy afterwards, and it is advised you take a family member or friend along who can take you home. A small dressing or plaster over the biopsy site can be removed the next day. There may be some mild bruising or discomfort, which usually is managed effectively by paracetamol. More serious complications such as bleeding or infection are very rare.

Lymph node biopsy

In some cases a lymph node (gland) biopsy is necessary to help confirm the diagnosis of CLL. This usually involves a small surgical procedure whereby an enlarged lymph node is removed. You will need a general anaesthetic for this and you will have a few stitches afterwards. Once the lymph node is removed it is examined in the laboratory by a pathologist. A pathologist is a doctor who is specially trained to examine tissue specimens and cells, to help diagnose diseases such as cancer.

Other tests

Once a diagnosis of CLL is made further tests may be done to find out the stage, or extent of the disease in your body and the effect it is having on other organs. They include a combination of blood tests and imaging tests. These tests can also provide important information about your general health and how well your kidneys, liver and other vital organs are functioning. The results may be important in selecting the best treatment for you. They can also be used as a baseline and compared with later results to assess how well you are progressing.

They may include:

Blood tests:

- kidney function
- liver function
- coagulation (to see if your blood is clotting properly)

Imaging tests:

- chest x-ray (to detect a chest infection or any other abnormalities)
- CT scan (to see if your spleen or liver is enlarged)

CT or CAT scan (computed axial tomography)

CT scans provide computer analysed, three dimensional (3D) images of cross sections of your body. This technology is able to detect tiny changes in tissue density which might indicate the presence of an infection or a tumour. The CT scan does not hurt and it usually takes less than an hour to complete. While the scan is being done you have to lie flat and still on a cushioned table that moves slowly through the CT machine. The machine itself looks like a giant ring surrounding the table. Sometimes a special contrast is used to enhance the quality of the pictures taken. The contrast may be swallowed or injected into a vein in your hand or arm before the scan. The CT scanner picks up the contrast as it moves through the body, highlighting areas to be examined more closely.

Waiting around for tests can be both stressful and boring. Remember to ask beforehand how long the test will take and what to expect afterwards. You might like to bring a book, some music, or a friend for company and support.



Staging and prognostic factors

Your doctor may want to document the stage of your disease and other prognostic factors that provide reliable information about the likely course of your disease (prognosis), and whether or not treatment should begin.

A prognosis is an estimate of the likely course of a disease. It provides some guide regarding the chances of curing the disease or controlling it for a given time. CLL is generally regarded as an incurable disease, but in many people the disease remains stable for long periods of time. If it does progress, effective treatment is available. The natural course of CLL can vary considerably between individuals. In around one third of cases people never require any treatment for their disease and may survive for many years without any effect from their disease. In another third CLL tends to progress gradually overtime, eventually requiring treatment. In the remainder of cases CLL presents as an aggressive disease, and needs to be treated soon after it is diagnosed.

Traditionally the Rai and Binet staging systems have been used to estimate prognosis in CLL. Using these systems patients are assigned to one of three major subgroups (good, intermediate or poor prognosis) depending on the number of lymphoid areas affected by the disease (lymph nodes, spleen or liver), and the red cell and platelet counts in the blood. Stage A refers to early disease, where in many cases people are asymptomatic and require no treatment. Stage B and C refer to more advanced disease which usually requires treatment.

Stage	Findings
Binet stage A / Rai stage 0	< 3 lymphoid areas involved Increased lymphocyte count
Binet stage B / Rai stage I and II	> 3 lymphoid areas involved Increased lymphocyte count
Binet stage C/ Rai stage III and IV	Increased lymphocyte count Low red cell count (anaemia) +/- low platelet count (thrombocytopenia)

Over recent years, there has been significant progress in identifying factors (known as prognostic factors), other than disease stage that provide additional reliable information about an individual patient's prognosis and how quickly their disease is likely to progress. These include the rate at which the leukaemic cells are multiplying. A fast (high) lymphocyte doubling time (less than 12 months) may be associated with a poorer prognosis for certain patients. The type of cytogenetic changes in affected lymphocytes also determines prognosis; some changes are favourable, others neutral and some such as 17p deletions are unfavourable. Changes in immunoglobulin genes (IgVH genes) affect prognosis. Lymphocytes normally undergo mutations of this area as they mature; unmutated immunoglobulin genes are associated with a less favourable prognosis. Because this test is difficult to do routinely other markers such as the cell surface marker CD38 and the protein ZAP-70 are

used to identify these patients. These factors may be used in deciding when to start treatment, especially in younger patients, and in patients with early stage disease.

Your doctor is the best person to give you an accurate prognosis regarding your disease, as he or she has the most information to make this assessment.

Commonly used response terms

The following terms may be used to describe how well your disease has responded to treatment.

Cure - This means that there is no evidence of leukaemia and no sign of it recurring, even after many years. This is not possible for most patients with CLL and the goal of current treatment is disease control.

Complete remission (CR) - This means that the treatment has been successful and that so much of the leukaemia has been destroyed that it can no longer be detected under the microscope. The proportion of blast cells in the marrow has been reduced to less than 5 per cent. There are no blast cells present in the circulating blood, the blood count has returned to normal, and no cytogenetic abnormalities can be detected. In CLL it also means when the bone marrow has regenerated, the proportion of lymphocytes is <30%.

Resistant/refractory disease - The leukaemia is not responding to treatment.

Relapse - The leukaemia has reappeared; it is no longer in remission

How is CLL treated?

The treatment chosen for your disease will depend on several factors including the stage of your disease, whether or not you have symptoms of your disease, how quickly your disease is progressing and your age and general health. Increasingly, treatment decisions are being influenced by other prognostic factors, described above, which may put some patients at a higher risk of disease progression than others regardless of the stage of their disease. The principle aims of treatment are two-fold: to prevent and reduce any symptoms of the disease and to bring about a long-lasting remission.

Information gathered from hundreds of other people around the world who have had the same disease helps to guide the doctor in recommending the best treatment for you.

Early stage CLL

Many people with CLL, particularly in the early stages of disease (Rai stage I and II, Binet stage 0) have no symptoms of their disease and don't require any treatment. Instead the doctor may recommend a 'watch and wait' strategy involving regular check-ups and blood counts to carefully monitor your health. This strategy may also be appropriate in more advanced stages of CLL, if your blood counts remain stable.

Advanced stage CLL

Treatment is usually only given when you start to have troubling symptoms of CLL, or when there are signs that your disease is starting to progress. Signs that your disease may be progressing include a significant increase in the number of lymphocytes in your blood (high lymphocyte doubling time), or rapidly growing lymph nodes.

There is a general agreement that most people with advanced stage CLL (Rai stage III and IV, Binet stage 2 and 3) need to be treated.

Remember that no two people are the same. In helping you to make the best treatment decision, your doctor will consider all the information available including the details of your particular situation.

Treatment for relapsed and resistant CLL

CLL is not usually curable and it usually comes back (relapses) after treatment. Often the relapse can be slow. Just because the disease has relapsed does not mean it needs to be treated, in fact, most people are observed for quite long periods before treatment needs to be restarted. Finding out that your leukaemia has relapsed or is resistant can be devastating, however there are usually several options for treating the disease and getting it back under control. The treatment of relapsed disease depends on a number of factors including the duration of the remission, the speed at which the disease has reappeared, and the number of times the leukaemia has been treated.

Your age and general health are also taken into account when considering which treatment is suitable for you.

Similar drugs to those used to initially treat your leukaemia or in some cases different drugs may be used to treat relapsed disease. You may also be invited to take part in a clinical trial to test new and experimental treatments for CLL. Younger patients who respond to chemotherapy for relapse may be considered for a transplant in some circumstances.

Palliative care

If a decision is made not to continue with anti-cancer treatment (chemotherapy etc) for your leukaemia there are still many things that can be done to help people to stay as healthy and comfortable as possible.

Palliative care is aimed at relieving any symptoms or pain a person might be experiencing as a result of their disease or its treatment, rather than trying to cure or control it.

Standard therapy

Standard therapy refers to a type of treatment which is commonly used in particular types and stages of disease. It has been tried and tested (in clinical trials) and has proven to be safe and effective in a given situation.

Informed consent

Giving your informed consent means that you understand and accept the risks and benefits of a proposed procedure or treatment. It means that you feel you have adequate information to make such a decision.

Your informed consent is also required if you agree to take part in a clinical trial, or if information is being collected about you or some aspect of your care (data collection).

If you have any doubts or questions regarding any proposed procedure or treatment, please do not hesitate to talk to the doctor or nurse again.

Types of treatment

Treatment for CLL may involve the use of:

- Chemotherapy
- Corticosteroid therapy
- Immunotherapy
- Radiotherapy
- Stem cell transplant
- Experimental treatments with drugs not yet available for general use (e.g. clinical trials)

Chemotherapy

Chemotherapy literally means therapy with chemicals. Many chemotherapy drugs are also called cytotoxics (cell toxic) because they kill cells, especially ones that multiply quickly like cancer cells.

Chemotherapy is the main form of treatment given for CLL. The dose, timing and types of the drugs used will vary depending on the particular disease involved, your age and general health, and the treatment protocol (plan of treatment) you are following.

Chemotherapy is usually given as a combination of drugs (combination chemotherapy). Each drug acts together to target the cancer in a different way, and also has different side effects. Therefore a combination may be more effective than a single drug in controlling your disease and the side effects are kept to a minimum. Chemotherapy is usually given in several cycles (or courses) with rest periods in between. This is to allow your body time to recover from the side effects.

Chemotherapy is given in many different ways in the treatment of CLL. Some drugs are given in tablet form (orally) or are injected into a vein (intravenously or IV) in your hand or arm.

In most cases you don't need to be admitted to hospital for chemotherapy, which is usually taken at home or given in the hospital's day treatment centre. Sometimes, depending on the type of chemotherapy being given and your general health, you may need to be admitted to the ward for a short while.

There are a variety of initial chemotherapy approaches. In general some approaches are better tolerated with lower response rates overall, but excellent responses in some individuals. Others, especially combination therapies, have more side effects but better overall response rates. An example of simple initial chemotherapy for CLL is a single oral (taken by mouth) drug like chlorambucil. An example of combination therapy is rituximab, cyclophosphamide and fludarabine. Studies have shown that both approaches work well. The choice of treatment for you depends on your and your doctors' preferences and the behaviour of your leukaemia cells.

The names of the different regimes used are commonly derived from the first letters of each of the drugs given. Some examples of combinations of drugs used to treat CLL are listed below.

FC	F ludarabine and C yclophosphamide
FCR	F ludarabine, C yclophosphamide and R ituximab (a monoclonal antibody)
CHOP	C yclophosphamide, H ydroxydaunorubicin (doxorubicin), O ncovin® (vincristine) and P rednisone (a corticosteroid)

Corticosteroid therapy

Corticosteroids are hormones produced naturally by the body. They can also be made in the laboratory. These drugs play an important role in the management of leukaemia. Prednisone and dexamethasone are examples of corticosteroids commonly used in the treatment of CLL. These drugs work by directly killing leukaemic cells as well as enhancing the effects of chemotherapy. They can also be useful in treating any autoimmune complication of CLL, such as haemolysis (red cell breakdown).

Immunotherapy

More recently, improved results have been achieved by combining chemotherapy with a monoclonal antibody like rituximab (MabThera) or alemtuzumab (CamPath). Monoclonal antibodies are specifically engineered to lock on to proteins found on the surface of abnormal cells like leukaemic cells.

Rituximab works by binding to the B-cell antigen CD20 found on the surface of the leukaemic cells. This helps the patient's own immune system to recognise these cells as foreign and kill them. Because this type of therapy specifically targets the leukaemic cells, they tend not to affect other healthy cells, which is why they are usually well tolerated with few side effects. Alemtuzumab works similarly, but targets the B-cell antigen CD52, it is generally well tolerated but is found on a greater number of other immune cells. There are a number of other antibody treatments being developed targeting the same or different antigens that may be offered as part of a research study.

Monoclonal antibodies are given as intravenous infusions, usually in the outpatient department of the hospital. Side effects are generally mild and can generally be easily managed. They may include fever, chills and mild skin reactions.

Radiotherapy

Radiotherapy is a type of treatment that uses high energy x-rays to kill cancer cells and shrink tumours. Radiotherapy is generally regarded as local therapy because it only destroys cancer cells in the treated area.

Radiotherapy is occasionally used in the treatment of CLL to shrink very enlarged lymph nodes or a very enlarged spleen which is causing symptoms. The spleen can also be surgically removed (splenectomy). The removal of the spleen in patients who have longstanding CLL may increase the patient's ability to have more treatment. In a small number of cases, radiotherapy is used in preparation for a stem cell transplant.

Before you start radiotherapy, the radiation oncologist (doctor who specialises in treating people with radiotherapy) will carefully calculate the correct dose of radiation therapy for you. The area or areas of your body that need to be treated will be marked with tiny ink dots on your skin using a special indelible pen.

Radiotherapy is usually given in small doses (also known as fractions) each weekday (Monday to Friday) over a week or more in the radiotherapy department of the hospital. You do not need to be admitted to hospital for this treatment, but if you live far away you may need to organise closer accommodation for this time. The social worker or nurses can assist you with this.

When you are having radiotherapy you usually lie on a table underneath the radiotherapy machine, which delivers the planned dose of radiation. If necessary, important structures like your heart and lungs are shielded as much as possible to ensure that they are not affected by the treatment given. Radiotherapy is painless. In fact you do not see or feel anything during the actual treatment. You will however need to stay perfectly still for a few minutes while the treatment is taking place. You might like to bring along some music to help you relax.

Stem cell transplant

A stem cell transplant will only be offered to you if your doctor feels that it will be of benefit. Transplants carry significant risks which your doctor will discuss with you if a transplant is a suitable treatment option in your case.

Allogeneic

Younger patients who have a suitably matched donor may be offered an allogeneic (donor) stem cell transplant when they have achieved their first remission from CLL. This involves the use of very high doses of chemotherapy, with or without radiotherapy, which kills the normal marrow cells (as well as, hopefully, any CLL cells that have survived thus far). The term used for this intense treatment is 'myeloablative'. This is then followed by an infusion of blood stem cells, which have been donated by another person; a suitably matched donor, usually a sibling or sometimes an unrelated donor from worldwide donor registries. Whether you will be offered a transplant will depend on a number of factors, predominantly the risk of relapse you are estimated to have if treated with chemotherapy alone together with your tolerability of the chemotherapy you will receive. This risk will vary between different patients so the advice from your doctor will be very specific to your circumstances. Due to the potential toxicities of this type of treatment it is not generally suitable for older patients (e.g. over 50 - 60 years).

An alternative approach involves using lower and therefore less toxic doses of chemotherapy and radiotherapy. This is called a reduced intensity, non-myeloablative, or mini-allogeneic (mini-allo) stem cell transplant. These reduced intensity transplants may also be suitable for selected older patients and those with certain health problems who would benefit from, but might not be able to tolerate a conventional donor transplant. Using this approach less intensive doses of chemotherapy are used to treat disease in the bone marrow and suppress the patient's immune system sufficiently for it to accept the new, donated healthy stem cells, leaving the donor's immune system to attack and destroy any leftover disease.

Autologous

Another option involves collecting your own stem cells, usually from your blood stream, storing them and then giving them back after you have received high doses of chemotherapy. This type of treatment is called an autologous stem cell transplant. This is commonly used for various forms of lymphoma and myeloma, but is very rarely used for CLL as studies have not shown it to be effective.

There are separate booklets about stem cell transplants available from Leukaemia & Blood Cancer New Zealand.

Common side effects

The type of side effects and their severity varies from person to person, depending on the type of treatment used and how an individual responds to it. There is no doubt that side effects can be very unpleasant at times, but it's good to remember that most of them are temporary and reversible.

It is important that you report any side effects you are experiencing to your nurse or doctor because many of them can be treated successfully, reducing any unnecessary discomfort for you.

It is important that you contact your doctor or the hospital for advice immediately (at any time of the day or night) if you are feeling very unwell, or if you experience any of the following:

- a temperature of 38°C or over and / or an episode of shivering
- bleeding or bruising, for example blood in your urine, faeces, sputum, bleeding gums or a persistent nose bleed
- nausea or vomiting that prevents you from eating or drinking or taking your normal medications
- diarrhoea, stomach cramps or constipation
- coughing or shortness of breath
- the presence of a new rash, reddening of the skin, itching
- a persistent headache
- a new pain or soreness anywhere
- if you cut or otherwise injure yourself
- if you notice pain, swelling, redness or pus anywhere on your body

Fatigue

Most people experience some degree of tiredness in the days and weeks following treatment for CLL. Having plenty of rest and a little light exercise each day may help to make you feel better during this time. Getting out into the fresh air and doing some gentle exercise is important for your general feeling of wellbeing and it also may help to reduce your fatigue. It is also important to listen to your body and rest when you are tired. Fatigue is a common side effect of CLL treatments and a symptom of CLL itself.

Autoimmune diseases

People with chronic lymphocytic leukaemia are more likely to develop autoimmune disease, due to impaired regulation of the immune system. The most common form of this is autoimmune red cell haemolysis, where you develop antibodies to your own red blood cells. The antibodies can be tested by doing a Coombs Test (also called a Direct Antibody test (DAT)). If you have this you may become jaundiced (yellow skin), have dark or orange urine, and become anaemic. It is important to tell your doctor if you develop any of these symptoms. Another common type of autoimmune problem is low platelets or ITP, this can cause easy bruising and petechiae (small pin prick bruises, described above) if the platelets are low enough. Other types of autoimmune diseases can occur but are less common e.g. Guillain-Barre Disease which causes nerve weakness.

Skin cancers

In New Zealand many people have sun damaged skin. Early skin cancers are normally kept in check by your immune system. In older patients this immune surveillance is not as efficient and squamous cell cancers (SCC), basal cell cancers (BCC) and less commonly melanomas can develop. The incidence of all of these cancers, especially SCC's and BCC's, is increased further in people with CLL as the immune surveillance is further impaired. In addition the skin cancers are often faster growing and more likely to spread. It is important that early skin cancers are treated, usually by your GP.

Effects on the bone marrow

CLL prevents your bone marrow from functioning properly and producing adequate numbers of red cells, white cells and platelets. Treatment, such as chemotherapy, also affects the bone marrow's ability to produce these cells. As a result, your blood count (the number of blood cells circulating in your blood) will generally fall within a week of treatment, increasing the risk of infection and bleeding.

Platelets -Your platelet count may also be affected by your disease and by the chemotherapy you are receiving and you may become thrombocytopenic (a reduction in the number of platelets circulating in the blood). When your platelet count is very low you can bruise and bleed more easily. During this time it is helpful to avoid sharp objects in your mouth such as toothpicks, as these can cut your gums. Using a soft toothbrush also helps to protect your gums. In many cases a transfusion of platelets is given to reduce the risk of bleeding until the platelet count recovers.

Red cells - If your red blood cell count and haemoglobin levels drop you will probably become anaemic. When you are anaemic you feel more tired and lethargic than usual. If your haemoglobin level is very low, your doctor may prescribe a blood transfusion.

White cells - The point at which your white blood cell count is at its lowest is called the nadir. This is usually expected 10 to 14 days after having your chemotherapy. During this time you will be at a higher risk of developing an infection. At this stage you will also be neutropenic, which means that your neutrophil count is low. Neutrophils are important white blood cells that help us to fight infection.

Sometimes your doctor may decide to use a growth factor such as G-CSF to help the recovery of your neutrophil count. This drug works by stimulating the bone marrow to increase the production of neutrophils. Your doctor and nurse will advise you on how to reduce your risk of infection while your white cell count is low.

Side effects of chemotherapy

Chemotherapy kills cells that multiply quickly, such as leukaemic cells. It also causes damage to fast-growing normal cells, including hair cells, and cells that make up the tissues in your mouth, gut and bone marrow. The side effects of chemotherapy occur as a result of this damage.

Chemotherapy in tablet form is tolerated well by most people and side effects tend to be few and mild. Intravenous chemotherapy can have more side effects, the type and severity of which will vary from one person to another, depending on what chemotherapy is used and how an individual responds to it.

There is no doubt that side effects can be very unpleasant at times but it's good to remember that most of them are temporary and reversible. It is important that you report any side effects you are experiencing to your nurse or doctor because many of them can be treated successfully, reducing any unnecessary discomfort for you.

Other side effects

Other possible side effects of chemotherapy less commonly seen in the treatment of CLL include:

- Nausea and vomiting
- Changes in taste and smell
- Mucositis (sore mouth)
- Diarrhoea or constipation
- Hair loss

Side effects of corticosteroids

The types of side effects seen with corticosteroids depend largely on how long they are used for, and the dose given. If you are using them for a short time you may notice that your appetite increases or you may feel more restless than usual. Some people find it more difficult to get to sleep at night and sleeping tablets or other natural therapies are sometimes recommended.

Corticosteroids can cause a rise in the blood sugar. Diabetics may find they need more of their diabetes medication while they are taking these drugs and some people who are not normally diabetic may require treatment to keep their blood sugar at acceptable levels. It is important to keep a check on the blood sugar and keep a diary of the levels and the amount of diabetic medication being taken. Diabetics will already know how to do this. People whose blood sugar only goes up when they are on corticosteroids may be given information on diet and taught how to measure their blood sugar and adjust their medication. Blood sugar levels usually return to normal once the steroids are finished.

Many of the side effects of corticosteroids are temporary and should pass once you finish taking them. Long-term use of corticosteroids may cause some other effects such as fluid retention and an increased susceptibility to infections. Aching joints such as the knees and hips have also been reported.

Remember to tell your doctor and nurses about any side effects you are having as they can usually suggest ways to help you.

Side effects of radiotherapy

Radiotherapy can cause similar side effects to those caused by chemotherapy including nausea and vomiting, hair loss and fatigue. These are described above.

In general the type of side effects seen with radiotherapy depends on the area of the body which has been treated. For example, radiotherapy to the abdomen is more likely to cause nausea and vomiting while hair loss is usually confined to the areas of the body being treated. It is not unusual to feel very tired after radiotherapy, so it is important to rest as much as you can during this time.

Skin reactions

Radiotherapy can cause a reddening of the skin which may also flake and become itchy. The staff at the radiotherapy department will advise you on how to care for your skin while you are having treatment. Gentle washing (avoiding perfumed products like scented soaps) and drying (patting rather than rubbing) is often recommended. You should also avoid any creams or moisturisers that contain traces of metals. Check with the radiotherapy department staff if you are unsure. Also, it is best to avoid direct sunlight on any area of skin that has received radiotherapy, even after the therapy has finished. This is because radiotherapy makes your skin more vulnerable to the damaging effects of the sun (i.e. sunburn and skin cancers).

Supportive care

Supportive care plays an important role in the treatment of many people with CLL. This involves making every effort to improve your quality of life, by relieving any symptoms you might have and by preventing and treating any complications that arise from your disease or treatment.

Blood transfusions, antibiotics, intravenous fluids and similar treatments, can all be important elements of medical supportive care.

Non-medical supportive care may involve complementary therapies, nutrition support, exercise, counselling and similar services.

Inform your haematologist if any surgery or treatment is planned by another practitioner, as advice may be required from your haematologist as to the best supportive treatment (such as transfusions, blood tests or other monitoring), to ensure that your treatment happens successfully without problems due to your disease, current or previous treatment.

Blood and platelet transfusions

If symptoms of anaemia are interfering with your normal daily activities, your doctor may recommend that you have a red blood cell transfusion. Platelet transfusions are sometimes given to prevent or treat bleeding (for example a persistent nose bleed). You do not need to be admitted to hospital for a red blood cell or platelet transfusion and they are usually given in the outpatient department. Transfusions don't usually cause any serious complications, nevertheless you will be carefully monitored throughout the transfusion. Alert your nurse if you are feeling hot, cold, and shivery or in any way unwell, as this might indicate that you are having a reaction to the transfusion. Steps can be taken to minimise these effects and ensure that they don't happen again.

Antibiotics

Infections are a common complication of CLL and its treatment, which can result for a number of reasons including lower levels of normally protective antibody (hypogammaglobulinaemia) and inadequate numbers of normally functioning white cells circulating in the blood stream. While infections can occur anywhere in the body; common sites include the upper and lower respiratory infections (chest infections), urinary tract (kidney infections) and skin. While most are caused by bacteria and viruses; fungal and opportunistic infections (infections caused by microorganisms that are normally harmless in healthy people) are also seen. You may be prescribed preventive (prophylactic) antibiotics especially during and after particular types of treatment. Some people are given monthly intravenous infusions of immunoglobulin to help fight infection.

While your white blood cell count is low you should take sensible precautions to help prevent infection. These include washing your hands frequently and using alcohol hand gel. Tell your friends and family, especially those with small children, that you are at risk of infection and ask them to ring before visiting if they have coughs and colds. Avoid crowds and other close contact with people who may have infections that are contagious (for example colds, flu, chicken pox). Only eat food that has been properly cooked and stored, and avoid sharing food and utensils.

If you do develop an infection you may experience a fever which may or may not be accompanied by an episode of shivering, where you shake uncontrollably. Infections while you are neutropenic can be quite serious and need to be treated with antibiotics as soon as possible.

It is important you do not use any drugs to bring your temperature down (i.e. paracetamol) until you are reviewed by your doctor. This could mask an infection which could lead to serious life threatening complications. Do not take aspirin or ibuprofen in any form as this can increase the risk of bleeding if your platelets are low. Always check with your doctor first.

Growth factors

As mentioned earlier, growth factors are natural chemicals in your blood that stimulate the bone marrow to produce different types of blood cells. Some of them can be made in the laboratory and used to help manage your CLL.

Erythropoietin (EPO) is an example of a growth factor which is used to stimulate the production of more red blood cells, and can in some cases reduce the need for frequent blood transfusions. Granulocyte-colony stimulating factor (G-CSF) may be given to stimulate the bone marrow to produce more white cells, particularly neutrophils. These white cells help fight bacterial and fungal infections in particular.

Growth factors are given as an injection under the skin (subcutaneous). They don't usually cause any major side effects but some people experience fevers, chills, headaches and some bone pain while using G-CSF. Your doctor may recommend that you take paracetamol to relieve any discomfort you may be feeling.

Complementary therapies

Complementary therapies are therapies which are not considered standard medical therapies. Many people find that they are helpful in coping with their treatment and recovery from disease. There are many different types of complementary therapies. These include yoga, exercise, meditation, prayer, acupuncture, relaxation and herbal and vitamin supplements.

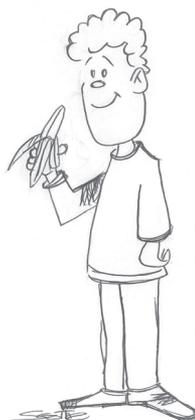
Complementary therapies should 'complement' or assist with recommended medical treatment. They are not recommended as an alternative to medical treatment. It is important to realise that no complementary or alternative treatment alone has proven to be effective against CLL.

It is also important to let your doctor or nurse know if you are using any complementary or alternative therapies in case they interfere with the effectiveness of chemotherapy or other treatments you may be having.

Nutrition

A healthy and nutritious diet is important in helping your body to cope with the condition you've been diagnosed with, and its treatment. Talk to your doctor or nurse if you have any questions about your diet or if you are considering making any radical changes to the way you eat. You may wish to see a nutritionist or dietician who can advise you on planning a balanced and nutritious diet.

If you are thinking about using herbs or vitamins it is very important to talk this over with your doctor first. Some of these substances can interfere with the effectiveness of chemotherapy or other treatment you are having.



Skin care and sun protection

Patients with CLL are at increased risk of developing skin cancer. It is important that you take care of your skin from the first day of CLL diagnosis with basic measures such as applying high factor sunscreen to exposed skin areas, and wearing long-sleeve clothes and a hat if you are heading outdoors. This is especially important if you are having chemotherapy and radiotherapy. Your doctor may refer you to a skin specialist (dermatologist) for regular check-ups.

Reproductive health

Fertility

Fertility is the ability to produce a child. In males, fertility means having enough healthy sperm to get a female pregnant. In females, fertility is the ability to become pregnant.

Some types of chemotherapy and radiotherapy may cause a temporary or permanent reduction in your fertility. It is very important that you discuss any questions or concerns you might have regarding your future fertility with your doctor if possible before you commence treatment.

In women, some types of chemotherapy and radiotherapy can cause varying degrees of damage to the normal functioning of the ovaries. In some cases this leads to menopause (change of life) earlier than expected. In men sperm production can be impaired for a while but the production of new sperm may become normal again in the future.

In younger patients there may be some options for preserving fertility, if this is an issue for you please discuss with your doctor if you believe you will be affected.

Early menopause

Some cancer treatments can affect the normal functioning of the ovaries. This can sometimes lead to infertility and an earlier than expected onset of menopause, even at a young age. The onset of menopause in these circumstances can be sudden and, understandably, very distressing.

Hormone changes can lead to many of the classic symptoms of menopause including menstrual changes, hot flushes, sweating, dry skin, vaginal dryness and itchiness, headache and other aches and pains. Some women experience decreased sexual drive, anxiety and even depressive symptoms during this time.

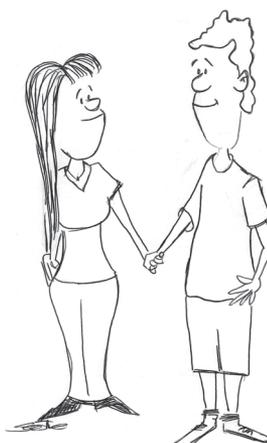
It is important that you discuss any changes to your periods with your doctor or nurse. He or she may be able to advise you or refer you to a specialist doctor (a gynaecologist) or clinic that can suggest appropriate steps to take to reduce your symptoms.

Body image, sexuality and sexual activity

It is likely that the diagnosis and treatment of leukaemia will have some impact on how you feel about yourself as a man or a woman and as a 'sexual being'. Hair loss, skin changes and fatigue can all interfere with feeling attractive.

During treatment you may experience a decrease in libido, which is your body's sexual urge or desire, sometimes without there being any obvious reason. It may take some time for things to return to 'normal'. It is perfectly reasonable and safe to have sex while you are on treatment or shortly afterwards, but there are some precautions you need to take.

It is usually recommended that you or your partner do not become pregnant as some of the treatments given might harm the developing baby. As such, you need to ensure that you or your partner uses a suitable form of contraception. Condoms (with a spermicidal gel) offer good contraceptive protection as well as protection against infection or irritation. Your partner may be worried that sex might in some way harm you. This is not likely as long as your partner is free from any infections and the sex is relatively gentle. Finally, if you are experiencing vaginal dryness, a lubricant can be helpful. This will help prevent irritation. Using a condom is also important to protect your partner from chemotherapy drugs that can be excreted in body fluids in the first few days after they are administered.



If you have any questions or concerns regarding sexual activity and contraception don't hesitate to discuss these with your doctor or nurse, or ask for a referral to a doctor or health professional who specialises in sexual issues.

Making treatment decisions

Many people feel overwhelmed when they are diagnosed with CLL. In addition to this, waiting for test results and then having to make decisions about proceeding with the recommended treatment can be very stressful. Some people do not feel that they have enough information to make such decisions while others feel overwhelmed by the amount of information they are given. It is important that you feel you have enough information about your illness and all of the treatment options available, so that you can make your own decisions about which treatment to have.



Anxiety, shock, denial or grief can make it difficult, at times, to absorb or remember discussions you have had with your doctor and it is common for people not to remember much of the information given to them at diagnosis. Before going to see your specialist doctor (haematologist) make a list of the questions you want to ask. It may be useful to keep a notebook with you and write questions down as you think of them, as often questions are forgotten between appointments.

Sometimes it is hard to remember everything the doctor has said. It may help to bring a family member or a friend along who can write down the answers to your questions or prompt you to ask others, be an extra set of ears or simply be there to support you.

Your doctor will spend time with you and your family discussing what he or she feels is the best option for you. Feel free to ask as many questions as you need to, at any stage. You are involved in making important decisions regarding your wellbeing.

You should feel that you have enough information to do this and that the decisions made are in your best interests. Remember, you can always request a second opinion if you feel this is necessary.

The Haematology Patient Diary, available from Leukaemia & Blood Cancer New Zealand, may be useful for recording details of treatment and making notes from clinic appointments.

Interpreting services

New Zealand's Health and Disability Code states that everyone has the right to have an interpreter present during a medical consultation. Family or friends may assist if you and your doctor do not speak the same language, but you can also ask your doctor to provide a trained interpreter if using a family member is not appropriate.

Social and emotional effects

People cope with a diagnosis of leukaemia in different ways, and there is no right or wrong or standard reaction. For some people, the diagnosis can trigger any number of emotional responses ranging from denial to devastation. It is not uncommon to feel angry, helpless and confused. Naturally people fear for their own lives or that of a loved one.

It is worth remembering that information can often help to take away the fear of the unknown. It is a good idea for you and your family to speak directly to your doctor regarding any questions you might have about your disease or treatment. It can also be helpful to talk to other health professionals including social workers or nurses who have been specially educated to take care of people with haematological diseases. Some people find it useful to talk with other patients and family members who understand the complexity of feelings and the kinds of issues that come up for people living with blood cancers and conditions.

In some areas there may be patient group meetings, and there is also an online support and information forum run by Leukaemia & Blood Cancer New Zealand – LifeBloodLIVE. This is available at www.lifebloodlive.org.nz.

Many people are concerned about the social and financial impact of the diagnosis and treatment on their families. Normal family routines are often disrupted and other members of the family may suddenly have to fulfil roles they are not familiar with, for example, cooking, cleaning, and taking care of children. The social worker attached to your treatment centre is available for you to talk through and discuss these issues with you and your family.

If you have a psychological or psychiatric condition, please inform your doctor and don't hesitate to request additional support from a mental health professional.

There is a variety of assistance available to help ease the emotional and financial strain created by a diagnosis of a blood cancer or condition. Support Services staff at Leukaemia & Blood Cancer New Zealand are available to provide you and your family with information and support to help you cope during this time. Contact details for Leukaemia & Blood Cancer New Zealand are provided on the back of this booklet.



Finishing treatment – Looking to the future

Once treatment has finished most people are followed up on a regular basis by their haematologist and are advised to see their general practitioner (GP) for any necessary medical care. This can make some people nervous because they may fear that their GP may not be aware of the latest developments in leukaemia. It is important to remember that your treating specialist will send information to your GP to keep him or her informed regarding your progress and what needs to be followed up on a regular basis, such as blood tests.

Even though you have been treated successfully for leukaemia it is normal to continue to experience feelings of vulnerability, uncertainty about the future and fear that your illness could return. The fear of a recurrence or relapse of leukaemia may cause some people to become over protective or cautious. Being more aware of any physical signs and symptoms than previously, for example a bruise, sustained in normal activity, may cause great anxiety and fear of relapse. Follow-up appointments after treatment has finished are often times of great anxiety as people wait for an 'all clear' from their doctor. As time passes and as more distance is allowed between appointments anxiety reduces. Everyone gradually becomes more and more engaged in the activities of daily living rather than concentrating most of their attention on the experience of illness.

Looking after yourself

Focusing on the things you can do to help yourself recover both physically and emotionally is important. Enjoying simple pleasures every day, looking to better times in the future, making plans and having hope are all important in maintaining a sense of control in a time of uncertainty.

Maintain a healthy lifestyle by:

- Avoiding smoking
- Eating a healthy diet
- Taking regular exercise
- Drinking alcohol in moderation
- Maintaining a healthy weight
- Wearing appropriate sun protection

Useful internet addresses

The value of the internet is widely recognised; however, not all the information available may be accurate and up to date. For this reason, we have selected some of the key sites that people with leukaemia might find useful.

With the exception of our own website, Leukaemia & Blood Cancer New Zealand do not maintain these listed sites. We have only suggested sites we believe may offer credible and responsible information, but we cannot guarantee the information on them is correct, up to date or evidence based medical information.

Leukaemia & Blood Cancer New Zealand

www.leukaemia.org.nz
www.lifebloodlive.org.nz

Cancer Society of New Zealand

www.cancernz.org.nz

Leukaemia Foundation of Australia

www.leukaemia.org.au

American Cancer Society

www.cancer.org

MacMillan Cancer Support (a UK cancer information site)

www.macmillan.org.uk

Leukemia & Lymphoma Society of America

www.leukemia-lymphoma.org

Leukaemia & Lymphoma Research Fund (UK)

www.llresearch.org.uk

National Cancer Institute (USA)

www.cancer.gov/cancerinfo

Grief Centre

www.griefcentre.org.nz

Glossary of terms

Alopecia

Hair loss. This is a side effect of some kinds of chemotherapy and radiotherapy. It is usually temporary.

Anaemia

A reduction in the haemoglobin level in the blood. Haemoglobin normally carries oxygen to all the body's tissues. Anaemia causes tiredness, paleness and sometimes shortness of breath.

Antiemetic

A drug which prevents or reduces feelings of sickness.

Blood count

Also called a full blood count (FBC). A routine blood test that measures the number and type of cells circulating in the blood.

B-lymphocyte

A type of white cell normally involved in the production of antibodies to combat infection.

Bone marrow

The tissue found at the centre of our bones. Active or red bone marrow contains stem cells from which all blood cells are made and in the adult this is found mainly in the bones making up the axial skeleton – hips, ribs, spine, skull and breastbone (sternum). The other bones contain inactive or (yellow) fatty marrow, which, as its name suggests, consists mostly of fat cells.

Cancer

A malignant disease characterised by uncontrolled growth, division, accumulation and invasion into other tissues of abnormal cells from the original site where the cancer started. Cancer cells can grow and multiply to the extent that they eventually form a lump or swelling. This is a mass of cancer cells known as a tumour. Not all tumours are due to cancer; in which case they are referred to as non-malignant or benign tumours.

Cannula

A plastic tube which can be inserted into a vein to allow fluid to enter the blood stream.

Central venous catheter (CVC)

Also known as a central venous access device (CVAD) or central venous line (CVL). A line or tube passed through the large veins of the arm, neck, chest or groin and into the central blood circulation. It can be used for taking samples of blood, giving intravenous fluids, blood, chemotherapy and other drugs without the need for repeated needles.

Chemotherapy

Single drugs or combinations of drugs which may be used to kill and prevent the growth and division of cancer cells. Although aimed at cancer cells, chemotherapy can also affect rapidly dividing normal cells and this is responsible for some common side effects including hair loss and a sore mouth (mucositis).

CT scan or CAT scan (Computerised axial tomography)

A specialised x-ray or imaging technique that produces a series of detailed three dimensional (3D) images of cross sections of the body.

Cure

This means that there is no evidence of disease and no sign of the disease reappearing, even many years later.

Echocardiogram

A special ultrasound scan of the heart.

Electrocardiogram (ECG)

Recording of the electrical activity of the heart.

Growth factors and cytokines

A complex family of proteins produced by the body to control the growth, division and maturation of blood cells by the bone marrow. Some are now available as drugs as a result of genetic engineering and may be used to stimulate normal blood cell production following chemotherapy or bone marrow or peripheral blood stem cell transplantation.

Haemopoiesis (or Haematopoiesis)

The processes involved in blood cell formation.

Haematologist

A doctor who specialises in the diagnosis and treatment of diseases of the blood, bone marrow and immune system.

Hairy cell leukaemia

A rare type of chronic leukaemia in which abnormal B-lymphocytes accumulate in the bone marrow, liver and spleen. Under the microscope, these cells are seen to have tiny hair-like projections on their surface.

High dose therapy

The use of higher than normal doses of chemotherapy to kill off resistant and / or residual (left over) cancer cells that have survived standard-dose therapy.

Immune system

The body's defence system against infection and disease.

Immunocompromised

When someone has decreased immune function.

Leukaemia

A cancer of the blood and bone marrow characterised by the widespread, uncontrolled production of large numbers of abnormal blood cells. These cells take over the bone marrow often causing a fall in blood counts. If they spill out into the bloodstream however they can cause very high abnormal white cell counts.

Leukaemic blasts

Abnormal blast cells which multiply in an uncontrolled manner, crowding out the bone marrow and preventing it from producing normal blood cells. These abnormal cells also spill out into the blood stream and can accumulate in other organs.

Leucopheresis

A procedure that uses a special machine called a 'cell separator' to remove the excess white cells in the blood while returning the rest of the blood to the patient.

Lymphocytes

Specialised white blood cells involved in defending the body against disease and infection. There are two types of lymphocytes, B-lymphocytes and T-lymphocytes. They are also called B-cells and T-cells.

Lymphomas

General name given to cancers of the lymphatic system.

Malignancy

A term applied to tumours characterised by uncontrolled growth and division of cells (see cancer).

Mucositis

Inflammation of the lining of the mouth and throat, which also can extend to the lining of the whole of the gastro-intestinal tract (stomach and intestines).

Neutropenia

A reduction in the number of circulating neutrophils, an important type of white blood cell. Neutropenia is associated with an increased risk of infection.

Neutrophils

Neutrophils are the most common type of white blood cell. They are needed to mount an effective fight against infection, especially bacteria and fungi.

Oncologist

General term used for a specialist doctor who treats cancer by different means, e.g. medical, radiation, surgical oncologist.

Pathologist

A doctor who specialises in the laboratory diagnosis of disease, and how disease is affecting the organs of the body.

Peripherally inserted central venous catheter (PICC)

A type of central venous catheter which is inserted into a large vein in the arm.

Prolymphocytic leukaemia (PLL)

A rare sub-type of chronic lymphocytic leukaemia in which abnormal lymphocytes known as prolymphocytes are found in the bone marrow and blood stream.

Prognosis

An estimate of the likely course of a disease.

Radiotherapy (radiation therapy)

The use of high energy x-rays to kill cancer cells and shrink tumours.

Resistant or refractory disease

This means that the disease is not responding to treatment.

Remission (or complete remission)

When there is no evidence of disease detectable in the body; note this is not always equivalent to a cure as relapse may still occur.

Spleen

An organ that accumulates lymphocytes, acts as a reservoir for red cells for emergencies, and destroys blood cells at the end of their lifespan. The spleen is found high in the abdomen on the left-hand side. It cannot normally be felt on examination unless it is enlarged. It is often enlarged in diseases of the blood – this is known as hypersplenism.

Splenomegaly

Another term used to describe an enlarged spleen.

Standard therapy

The most effective and safest therapy currently being used.

Stem cells

Stem cells are primitive blood cells that can give rise to more than one cell type. There are many different types of stem cell in the body. Bone marrow stem cells have the ability to grow and produce all the different blood cells including red cells, white cells and platelets.

Stem cell transplant (peripheral blood stem cell or bone marrow transplant)

These treatments are used to support the use of high-dose chemotherapy and / or radiotherapy in the treatment of a wide range of blood cancers including leukaemias, lymphomas, myeloma, certain solid tumours, and other serious diseases.

T-lymphocyte

A type of white cell involved in controlling immune reactions.

Tumour

An abnormal mass of cells which may be non-malignant (benign) or malignant (cancerous).

Ultrasound

Pictures of the body's internal organs built up from the interpretation of reflected sound waves.

White blood cells (White cells)

Specialised cells of the immune system that protect the body against infection. There are five main types of white blood cells: neutrophils, eosinophils, basophils, monocytes and lymphocytes.

X-ray

A form of electronically produced radiation used in diagnosis and treatment.

Please refer to the 'Dictionary of Terms' booklet for further definitions.

Please send me a copy of the following patient information booklets:

- | | |
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| <input type="checkbox"/> Dictionary of Terms | <input type="checkbox"/> Acute Lymphoblastic Leukaemia in Adults |
| <input type="checkbox"/> Haematology Patient Diary | <input type="checkbox"/> Acute Lymphoblastic Leukaemia in Children |
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Address: _____

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Send to: Leukaemia & Blood Cancer New Zealand
 PO Box 99182, Newmarket, Auckland 1149
 Phone: 09 638 3556 or 0800 15 10 15
 Email: info@leukaemia.org.nz

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Chronic Lymphocytic Leukaemia

We hope that you found this information booklet useful. We are interested in what you thought of the booklet – whether you found it helpful or not. If you would like to give us your feedback, please fill out this questionnaire and send it to Leukaemia & Blood Cancer New Zealand, at the address at the bottom of the following page.

1. Did you find this booklet helpful?

- Yes No

Comments _____

2. Did you find this booklet easy to understand?

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3. Where did you get this booklet from?

4. Did you have any questions that were not answered in the booklet?

- Yes No

If yes, what were they?



5. What did you like the most about this booklet?

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7. Any other comments?

Thank you for helping us review this booklet. We will record your feedback and consider it when this booklet is reviewed for the next edition.

Please return to: Leukaemia & Blood Cancer New Zealand
PO Box 99182 Newmarket, Auckland 1149



Important information for haematology patients

It is important that you contact your doctor or the hospital for advice immediately (at any time of the day or night) if you are feeling very unwell, or if you experience any of the following:

- a temperature of 38°C or over and / or an episode of shivering
- bleeding or bruising, for example blood in your urine, faeces, sputum, bleeding gums or a persistent nose bleed
- nausea or vomiting that prevents you from eating or drinking or taking your normal medications
- diarrhoea, stomach cramps or constipation
- coughing or shortness of breath
- the presence of a new rash, reddening of the skin, itching
- a persistent headache
- a new pain or soreness anywhere
- if you cut or otherwise injure yourself
- if you notice pain, swelling, redness or pus anywhere on your body



our mission is to care, our vision is to cure

Freephone 0800 15 10 15
Telephone 09 638 3556
Facsimile 09 638 3557
Email info@leukaemia.org.nz

National Office:

6 Claude Road, Epsom 1023
PO Box 99182, Newmarket 1149
Auckland, New Zealand

leukaemia.org.nz

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