Acute Lymphoblastic Leukaemia (ALL) in Adults

A guide for patients, families & whānau

leukaemia & blood cancer new zealand

our mission is to care, our vision is to cure

Contents

There is a separate information booklet called 'Acute Lymphoblastic Leukaemia in Children – a guide for parents, families and whānau' available from Leukaemia & Blood Cancer New Zealand.

Leukaemia & Blood Cancer New Zealand is grateful to the Newmarket Rotary Charitable Foundation for sponsoring this booklet

Introduction

This booklet has been written to help you and your family or whānau understand more about acute lymphoblastic leukaemia (ALL) in adults.

If you or someone you care for has been diagnosed with ALL, 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 Richard Doocey (Auckland City Hospital) and Dr Liam Fernyhough (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.

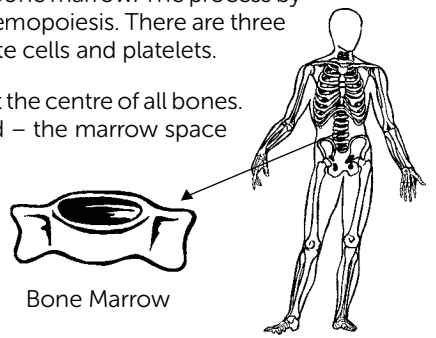
We welcome visitors to our offices in Auckland, Wellington and Christchurch. Please phone for an appointment.

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). Some of you may have had a bone marrow biopsy taken from the bone at the back of your hip (the iliac crest) or the breastbone.

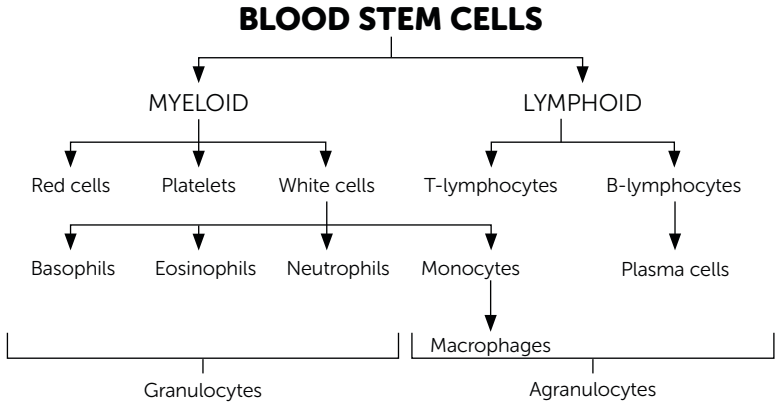


Bone Marrow

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.

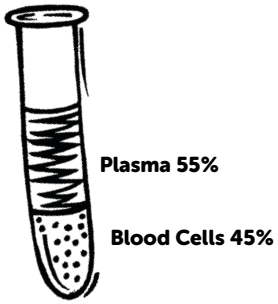
Myeloid ('my-loid") stem cells develop into red cells, white cells (neutropils, eosinophils, basophils and monocytes) and platelets.

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 count is between 4.0 – 11.0 x 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 x10^9/L) you are considered to be neutropenic and at risk of developing frequent and sometimes severe infections.

The normal adult neutrophil count is between 2.0 – 7.5 x 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 count is between 150 - 400 x 10^9/L

Thrombocytopenia is the term used to describe a reduction in the normal 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 x 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

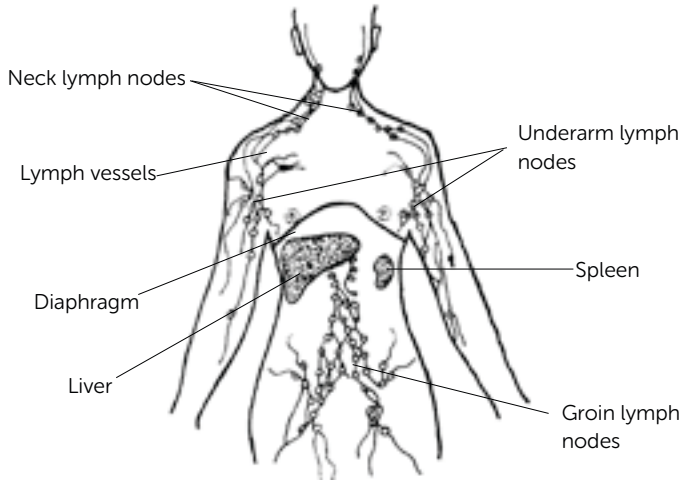
All normal blood cells have a limited survival in the circulation and need to be replaced on a continual basis. This means that the bone marrow remains a very active tissue throughout your life. Natural chemicals in your blood called growth factors or cytokines control the process of blood cell formation. 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 acute lymphoblastic leukaemia (ALL)?

Acute lymphoblastic leukaemia (ALL) is a type of cancer that affects immature lymphocytes developing in the bone marrow. Under normal condition these cells grow and mature into specialised white cells called B-lymphocytes (B-cells) and T-lymphocytes (T-cells). In ALL, they multiply in an uncontrolled way, quickly crowding the bone marrow, and interfering with normal blood cell production. Because the bone marrow is unable to make adequate numbers of red cells, normal white cells and platelets, people with ALL become more susceptible to anaemia, recurrent infections and to bruising and bleeding easily.

Excessive numbers of these abnormal lymphocytes, known as lymphoblasts, leukaemic blasts or leukaemic cells, spill out of the bone marrow and circulate around the body in the bloodstream. From here they can accumulate in various organs including the lymph nodes (glands), spleen, liver and central nervous system (brain and spinal cord).

How common is ALL and who gets it?

Each year in New Zealand around around 600 adults and 40 children are diagnosed with leukaemia. ALL is the most common type of childhood leukaemia, while in adults acute myeloid leukaemia (AML) is more commonly diagnosed. The percentage of all acute leukaemias due to ALL falls with age; ALL is more common in males than in females.

Overall, chronic leukaemia is more common in adults than acute leukaemia.

The characteristics of ALL differ greatly between children and adults. Current treatment regimes mean that the majority of children with ALL successfully treated. In adults, cure rates are more variable.

What causes ALL?

Many people who are diagnosed with ALL 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 ALL remains unknown. We do know that it is not contagious, that is, you cannot 'catch' ALL by being in contact with someone who has it.

Like other types of leukaemia, ALL 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, exposure to very large doses of radiation, or certain drugs used to treat other forms of cancer may increase the risk of ALL. Certain types of viral infections may play a role in the development of some types of ALL.

What are the symptoms of ALL?

Because ALL develops quickly, people are usually only unwell for only a short period of time before they are diagnosed (days or weeks). The most common symptoms of ALL are caused by a shortage of normal blood cells in the circulating blood. These include:

Anaemia

A low haemoglobin level in the blood can cause symptoms of anaemia. These include lack of energy, persistent tiredness and fatigue, weakness, dizziness or feeling unusually short of breath when physically active. In addition, people with anaemia often have a pale complexion.

Increased bleeding or bruising

A very low platelet count can cause bruising for no apparent reason, or excessive or prolonged bleeding following minor cuts or injury. Some people notice frequent or severe nose bleeds or bleeding gums and some women may have unusually heavy menstrual periods. Red or purple flat pinhead sized purple spots may appear on the skin, especially on the legs. These are called petechiae ('pe-tee-kee-i') and they are caused by tiny bleeds under the skin.

Frequent or repeated infections

People with ALL don't have enough normal white blood cells, particularly neutrophils, so they are more likely to develop frequent or repeated infections. These may present as minor skin infections, slow healing of minor cuts and grazes, a sore throat, sore mouth, coughing, urinary tract infections (frequent passing of urine with a sensation of burning) and often fevers.

Bone pain

Pain in the bones and joints is common and results from the marrow being crowded with leukaemic cells.

Other symptoms of ALL may include swollen lymph nodes (glands), chest pain and abdominal discomfort due to a swollen spleen or liver. Some people, particularly those with T-cell ALL may experience chest pain and shortness of breath due to swollen lymph nodes in the chest. This is known as a mediastinal mass. Occasionally, leukaemic cells can accumulate in the skin causing a rash.

Some of the symptoms described above may also be seen in other illnesses, including viral infections. 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 appropriately.

Which doctor?

If your GP suspects that you might have leukaemia 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.

How is ALL diagnosed?

ALL is diagnosed by examining samples of your blood and bone marrow.

Full blood count

The first step in diagnosing ALL 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 hand or 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 ALL.

Many people with ALL have a low red cell count, a low haemoglobin level (anaemia) and a low platelet count. Most will also have a high white cell count with large numbers of abnormal leukaemic blast cells (lymphoblasts) in the circulating blood The presence of leukaemic cells in the bloodstream suggests that you have leukaemia. A small percentage of patients may not have lymphoblasts detected in their blood at diagnosis. In all cases, the diagnosis will need to be confirmed by examining the cells in your bone marrow.

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

Bone marrow examination

A bone marrow examination (or biopsy) 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. A diagnosis of ALL is confirmed by the presence of an excessive number of blast cells in the bone marrow. Under normal circumstances the bone marrow contains a small proportion of normal or healthy blast cells, usually less than 5 per cent. This proportion can increase to between 20% and 95% in people with ALL.

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.

Once a diagnosis of ALL is made, blood and bone marrow cells are examined further using special laboratory tests. These include immunophenotyping, cytogenetic and molecular tests.

These tests provide more information about the exact type of disease, the likely course of the disease and the best way to treat it.

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

Immunophenotyping looks at special markers called antigens found on the surface of blast cells to determine the exact subtype of leukaemia and therefore the best way to treat it. This test is done on a machine called a flow cytometer and the test is often called flow cytometry. Specific patterns of antigens on leukaemia cells can be used to follow the leukaemia and check how well it is responding.

Antigens, commonly referred to as 'cluster of differentiation' or CD antigens followed by a number, act like flags identifying the type and origin of a cell and distinguishing it from other cells in a given sample. Recognition of particular CD antigens is useful in distinguishing between normal and leukaemic cells and determining the type of cell in which the leukaemia originated (B-lymphocyte – B-cell ALL or T-lymphocyte – T-cell ALL), and the point at which this cell stopped developing properly in the bone marrow.

Cytogenetic ('cy-to-gen-etic') and molecular genetic tests

Cytogenetic tests provide information about the genetic make-up of the leukaemic cells, in other words, the structure and number of chromosomes present. Chromosomes are the structures that carry genes. Genes are collections of DNA, our body's blueprint for life. Standard cytogenetic tests involve examining the chromosomes under the microscope.

Chromosome changes

Certain cytogenetic changes, such as missing, extra or abnormal chromosomes help to confirm the specific sub-type of ALL you have, and which treatment is likely to be most effective. These chromosomal changes are only found in the leukaemic cells. They are not usually passed down from parent to child (inherited). Instead, they tend to be acquired over time. An example of this is the Philadelphia (Ph) chromosome, found in some leukaemic cells. This abnormal chromosome is formed when part of chromosome 9 (the ABL gene) breaks off and attaches itself to part of chromosome 22 (the BCR gene) in a process known as translocation. This translocation t(9;22) produces the new fusion gene BCR-ABL which in turn releases excess amounts of an enzyme called tyrosine kinase. Tyrosine kinase continually signals the bone marrow to make too many abnormal blood cells.

The Ph chromosome is the most common chromosomal abnormality seen in adults with ALL (B cell), occurring in 25 to 30 per cent of all adult patients. Its frequency increases with age and is as high as 50 per cent in people over the age of 50 years. Ph chromosome positive (Ph+) ALL tended to respond poorly to conventional chemotherapy but newer oral drugs called tyrosine kinase inhibitors (for example imatinib mesylate or dasatinib) are now used in combination with chemotherapy and have substantially improved the response rate. In suitable patients, an allogeneic (donor) stem cell transplant may be considered at an earlier stage.

Molecular genetic tests (for example polymerase chain reaction or PCR tests and fluorescent in situ hybridization or FISH) are more sophisticated genetic tests which may be used to assess how well your disease has responded to treatment. These tests are capable of measuring minute traces of leftover (residual) leukaemic cells not normally visible under the microscope. The presence of left over disease gives the doctor some indication of the likelihood of future relapse (return of the original disease). Using this highly sensitive technology, subtle changes in your disease can be detected earlier and where necessary treated earlier.

Together, immunophenotyping, cytogenetic and molecular tests provide more information about the exact type of disease you have, it's likely response to treatment and the best way to treat it.

Other tests

Other tests provide information on your general health and how well your kidneys, liver and other vital organs are functioning. These include a combination of blood tests and x-rays. Blood tests may include kidney function tests, liver function tests and coagulation tests, to see if your blood is clotting properly.

A lumbar puncture is a procedure where a small sample of the cerebro-spinal fluid (CSF) that surrounds your brain and spinal cord is collected via a needle in the lower back. This fluid is tested in the laboratory to check for the presence of leukaemic cells within the central nervous system.

These tests are important because they provide a baseline set of results regarding organs that might be affected by disease, and your general health. The results may be important in selecting the best treatment for you. The results can also be compared with later results to assess how well you are progressing.

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.

Which type of ALL do I have?

ALL is not a single disease. It is the name given to a group of leukaemias that develop in the lymphoid cell line in the bone marrow. Depending on the type of abnormal lymphocyte present, ALL can be broadly classified into two main groups:

- ALL that arises in developing B-lymphocytes (B-cells)
- ALL that arises in developing T-lymphocytes (T-cells)

The current World Health Organization's (WHO) classification system for ALL uses additional information, obtained from more specialised laboratory techniques, like immunophenotyping and cytogenetic tests (see above), to classify ALL precisely. The diagnosis of different subtypes of ALL depends on the presence or absence of distinct cell surface markers (CD antigens; see page 14).

Pre-B-cell ALL

Between 75 to 80 per cent of adult ALL arises in B-lymphocytes in the early stages of development in the bone marrow. In these cases the affected cells share several characteristics with normal immature B-cells. The disease is therefore called precursor B-cell ALL or Pre-B-cell ALL.

B-cell ALL

B-cell ALL arises in more mature developing lymphocytes. This type of ALL is less common accounting for around 3 to 5 per cent of all adult cases.

Here leukaemic cells tend to spread to areas outside the blood and bone marrow and collections of lymphoblasts can be found in areas such as the abdomen, head and neck regions. Involvement of the central nervous system is common.

B-cell ALL sometimes resembles another disease called Burkitts lymphoma, a rare aggressive type of lymphoma. People diagnosed with B-cell ALL are commonly treated with drugs similar to those used to treat this lymphoma.

T-cell ALL

In around 20 to 25 per cent of cases ALL arises in developing T-cells. This type of ALL can be further classified as early, mid or late thymocyte T-cell ALL, depending on the maturity of the affected cell. T-cell ALL commonly presents with a high white blood cell count and may involve the central nervous system at diagnosis. In many cases, leukaemia has spread to the chest where swollen lymph nodes produce a collection known as a mediastinal mass.

Prognosis and treatment differ between adults and children. There is a separate information booklet called 'Acute Lymphoblastic Leukaemia in Children – a guide for parents, families and whānau' available from Leukaemia & Blood Cancer New Zealand.

Prognosis

A prognosis is an estimate of the likely course of a disease and whether it is likely to relapse in the future. It provides some guide regarding the chances of curing the disease or controlling the disease for a given time.

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

Certain factors (known as prognostic factors) give some patients a better chance of being cured of their disease with treatment than others. At the start these include your age, the exact type of disease you have and your white cell count at diagnosis. The genetic make-up of the leukaemic cells is an important factor in predicting prognosis and the likelihood of cure in ALL. For example, certain cytogenetic changes are associated with a less favourable, prognosis than others. The presence of Ph+ disease, or leukaemic cells with less than the normal number of chromosomes present (hypodiploidy), have historically been associated with a poorer prognosis using standard therapy (although, as mentioned, this may no longer apply to Ph+ ALL).

Another important prognostic factor is how well your disease responds to initial treatment, that is, how quickly you achieve a remission and how much disease is left over in your body after this initial treatment.

Taking these and other factors into consideration, you will be categorised as having 'standard-risk' or 'high-risk' ALL. This ensures that the most appropriate and effective 'risk-based' therapy can be chosen for you. For example, more intensive therapy may be more beneficial than standard therapy for some people who are in the high-risk group. Intensive therapy may help to reduce your risk of future relapse and therefore increase your overall chances of survival.

It is important to realise that although the majority of people treated for ALL will achieve a remission, a significant proportion will relapse over time. It is currently estimated that, with chemotherapy treatment alone, overall between 20 to 40 per cent of adults can be cured of ALL.

Commonly used prognostic 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 re-appearing, even after many years.

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. The term commonly used for these tests is measuring the level of 'minimal residual disease' or MRD.

The length of time that a remission lasts may vary from person to person, and the leukaemia may well reappear (relapse) over time.

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

Relapse - The leukaemia has reappeared.

How is ALL treated?

The treatment chosen for your disease depends on a number of factors including the exact type of leukaemia you have, your age, other prognostic factors, and your general health.

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.

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.

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.

Clinical trials

Your specialist doctor may ask you to consider taking part in a clinical trial (also called a research study). Clinical trials test new treatments, or existing treatments given in new ways to see if they work better. Clinical trials are important because they provide vital information about how to improve treatment by achieving better results with fewer side effects.

Participation in a trial may also involve giving blood or bone marrow samples in order to contribute to a better understanding of the disease. Clinical trials often give people access to new therapies not yet funded by governments.

Taking part in a clinical trial is entirely voluntary and you are under no obligation to participate. If you are considering taking part in a clinical trial, make sure that you understand the reasons for the trial and what it involves for you. You should always take time to consider all the implications of a trial and discuss this thoroughly with your specialist doctor and other support people before giving your informed consent. Your specialist doctor can guide you in making the best decision for you.

There is a separate booklet called 'Clinical Trials' available from Leukaemia & Blood Cancer New Zealand.

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

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 ALL. 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). These drugs act together and in different ways to destroy the leukaemic cells. 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 ALL. Some drugs are given in tablet form (orally) but most are injected into a vein (intravenously or IV). IV drugs are usually given through a special line called a central venous catheter (or central line). This is a special line inserted through the skin, into a large vein in your arm, neck or chest. Once in place, chemotherapy and other drugs can be given through the line. There are several different kinds of central lines used; some are intended for short-term use while others can remain in place for months or even years.

Cortico-steroid therapy

Cortico-steroids 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 cortico-steroids commonly used in the treatment of ALL. These drugs work by directly killing leukaemic cells as well as enhancing the effects of chemotherapy.

Central nervous system (CNS) treatment and prophylaxis

Leukaemic cells are sometimes found in the CNS (brain and spinal cord) at the time of diagnosis. In other cases ALL reappears or relapses within this area at a later stage. Because the blood supply to the CNS is different from the blood supply to other parts of the body, this area can act as a 'sanctuary site' or hiding spot for leukaemic cells. Here the cells can grow and multiply beyond the reach of standard chemotherapy drugs which normally travel throughout the rest of the body in the blood stream.

CNS treatment and prophylaxis (protection) will be given at various stages throughout your treatment. This usually involves injections of methotrexate and / or other chemotherapy drugs directly into the spinal fluid (intrathecal injection), through a lumbar puncture. Some types of intravenous chemotherapy and cortico-steroid therapy also provide valuable protection for the CNS. On rare occasions, radiation therapy to the head (cranial irradiation) is also used.

New treatments for ALL

There are several new and promising approaches under development for the treatment of ALL. These include new chemotherapy drugs such as clofarabine and nelarabine and existing chemotherapy drugs such as vincristine and daunorubicin which have been encapsulated in a liposomal (fat) solution. Liposomal preparations may allow higher total doses of chemotherapy to be given without causing an increase in toxicity to normal cells.

Newer targeted therapies are also being developed including second-generation tyrosine kinase inhibitors (eg dasatinib or a similar drug called nilotinib) for Ph+ disease and monoclonal antibodies (rituximab and alemtuzumab). Monoclonal antibodies are specifically engineered to lock on to different proteins, called antigens, found on the surface of abnormal cells like leukaemic cells. Rituximab binds to CD20 antigens and studies from overseas have shown that giving rituximab improves the outcome for patients whose ALL has the CD20 antigen on the surface of the leukaemia cells. In contrast, alemtuzumab binds to a CD52 antigen. 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 may explain why they are usually well tolerated with few side effects.

Most of these new treatments for ALL are not freely available but maybe available as part of a clinical trial in some centres or may be available under a compassionate access scheme. Your doctor will be able to discuss with you all of the treatment options suitable for you.

ALL in adolescents and young adults

Recent studies suggest that adolescents and young adults may have better outcomes using paediatric treatment protocols which traditionally have been more intensive than adult protocols. Trials are currently under consideration to determine if these dose-intensive protocols could also improve outcomes for adults aged between 18-35 years.

Phases of treatment

Treatment for ALL can be divided into three phases:

- Induction
- Consolidation
- Maintenance

Induction

Soon after you are diagnosed you will start an intensive course of treatment to bring about, or induce, a remission. The goal of this treatment is to destroy any detectable leukaemic cells in your blood and bone marrow and allow your bone marrow to function normally again. You will need to be admitted to hospital for this first phase of treatment.

Commonly used chemotherapy drugs in this phase of treatment include: vincristine, cyclophosphamide and an anthracycline drug (daunorubicin, adriamycin). Other drugs like cortico-steroids (prednisone) are also used. CNS therapy also may begin at this stage. Another important drug in ALL treatment both in induction and later on is asparaginase which through enzyme activity leads to leukaemia cell death.

While you are having induction therapy you may also be given a drug called allopurinol, which is not a chemotherapy drug. It is used to help prevent a build-up of breakdown products of the destroyed leukaemic cells and to help the kidneys excrete these products safely. High volumes of fluid are also given intravenously to help flush through the kidneys. In patients where there is a high risk of this complication (such as very high leukaemia cell count) a drug called rasburicase may be used to protect the kidneys.

Usually the administration of induction chemotherapy and the recovery of the bone marrow from this treatment takes about 4 weeks. At the end of this time you will undergo another bone marrow biopsy and testing to assess whether you are in remission or not. The tests include microscopic examination, cytogenetics and, in some patients where applicable, flow cytometry and molecular tests

The majority of patients (around 80 per cent) will achieve an initial remission following induction therapy. In a small number however the disease does not respond to treatment as expected (e.g. the blasts cell count in your marrow does not normalise or flow cytometry or molecular tests suggest there is significant residual leukaemia) and you may be said to have resistant or refractory disease. In these cases the doctor may recommend a more intensive form of therapy to treat your disease more effectively.

People with Ph+ disease may also be treated with a drug called imatinib mesylate (imatinib), a tyrosine kinase inhibitor, during the induction and post-remission phases of their treatment. Imatinib is a relatively new drug that works by targeting the bcr-abl protein thereby blocking the leukaemia-causing effects of tyrosine kinase. Higher remission rates have been reported using a combination of imatinib and chemotherapy although the effects on long-term survival are as yet unclear. Newer research protocols are using an alternate tyrosine kinase inhibitor called dasatinib instead of imatinib.

Consolidation

Soon after remission induction therapy finishes, more treatment is required to destroy any left over disease as just one cycle of chemotherapy is rarely sufficient to get rid of all the leukaemia cells, even in those patients who achieve remission. This is important because it helps to prevent the disease from reappearing (relapsing) or spreading to the central nervous system (brain and spinal cord) in the future. This second phase of treatment is called consolidation, post remission therapy, or intensification. The type of consolidation therapy chosen for you will depend on your estimated risk of relapse in the future, in other words the 'risk group' to which you belong (see below). Consolidation therapy usually involves 'blocks' of treatment over several months. This often includes several courses of more intensive chemotherapy (intensification) to eradicate residual disease.

Because of the high risk that ALL will relapse in the future, some people may be offered even more intensive treatment followed by a stem cell transplant, to more effectively treat their disease.

Maintenance

Maintenance therapy is designed to help keep your disease in remission and prevent it from reappearing (relapsing) in the future. Common maintenance protocols involve chemotherapy tablets, some taken daily, others weekly, and possibly blocks of injections of chemotherapy with courses of cortico-steroids.

This phase of treatment usually lasts from several months to up to 2 years during which time you will be treated as an outpatient. However, depending on the type of chemotherapy being given or your general health, you may need to be admitted to hospital.

Stem cell transplant

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 ALL. 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 ALL cells that have survived thus far). The term used for this intense treatment is 'myeloablative'. This is then followed by infusion of blood stem cells, which have been donated by a suitably matched donor, usually a sibling or sometimes an unrelated donor from worldwide donor registries. This form of treatment may reduce the chance of relapse in both standard and high-risk ALL and improve the overall chance of cure. Whether you will be offered a transplant will depend on a number of factors, predominantly the risk of relapse if treated with chemotherapy alone together with how you tolerate 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).

A newer 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. This may 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. Meanwhile it is hoped that the donor's immune system will 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 ALL as studies have not shown it to be effective.

A stem cell transplant is usually only offered if your doctor feels that it will be of benefit to you. You will be able to discuss with your doctor if a transplant is a suitable treatment option in your case.

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

Relapsed disease

Finding out that your leukaemia has relapsed can be devastating, but there are usually ways of getting it back under control. The treatment of relapsed disease depends on a number of factors including the duration of the remission and the site at which the disease has reappeared. Other factors are also considered including your age and the genetic make-up of the relapsed leukaemic cells.

Similar drugs to those used to initially treat 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 ALL. 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 and radiotherapy) 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.

Common side effects

The type of side effects and their severity varies from person to person, depending on the type of chemotherapy 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

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.

Effects on the bone marrow

ALL prevents your bone marrow from functioning properly and producing adequate numbers of red cells, white cells and platelets. 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 their 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.

While your white blood cell count is low you should take sensible precautions to help prevent infection. These include avoiding crowds, avoiding close contact with people with infections that are contagious (for example colds, flu, chicken pox) and only eating food that has been properly prepared and cooked.

Your doctor and nurse will advise you on how to reduce your risk of infection while your white cell count is low.

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 you doctor first.

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. G-CSF is given as an injection under the skin (subcutaneous). This is quite a simple procedure and the nurse will teach you or a family member (or friend) to do this at home. Major side effects are uncommon, but occasionally aching bones may occur.

Nausea and vomiting

Nausea and vomiting are often associated with chemotherapy and some forms of radiotherapy. However, thanks to significant improvements in anti-sickness (antiemetic) drugs, nausea and vomiting are generally very well controlled. You will be given anti-sickness drugs before and for a few days after your chemotherapy treatment. Be sure to tell the nurses and doctors if you think that the antiemetics are not working for you and you still feel sick. There are many different types of antiemetics that can be tried. A mild sedative may also be used to help stop you feeling sick. This will help you to relax but it might make you a little sleepy.

Some people find that eating smaller meals more frequently during the day, rather than a few large meals, helps to reduce nausea and vomiting. Many find that eating cool or cold food is more palatable, for example jelly or custard. Drinking ginger ale or soda water and eating dry toast may also help if you are feeling sick. Getting plenty of fresh air, avoiding strong or offensive smells and taking the prescribed anti-sickness drugs as recommended by the nurse and doctor should also help.

Changes in taste and smell

Both chemotherapy and radiation therapy can cause changes to your sense of taste and smell. This is usually temporary but in some cases it lasts up to several months. During this time you may not be able to enjoy the foods and drinks that you used to love and this can be very disappointing. Some people find that adding a little more sugar to sweet foods and salt to savoury foods can help. Talk to your dietician for advice.

Mucositis

Mucositis, or inflammation of the lining of the mouth and throat, is a common and uncomfortable side effect of chemotherapy and some forms of radiotherapy. It usually starts about a week after the treatment has finished and goes away once your blood count recovers, usually a couple of weeks later. During this time your mouth and throat could get quite sore. Soluble paracetamol and other topical drugs (ones which can be applied to the sore area) can help. If the pain becomes more severe, stronger pain killers might be needed.

Always check your temperature before taking paracetamol as this drug can 'mask' signs of infection (a raised temperature).

It is important to keep your mouth as clean as possible while you are having treatment to help prevent infection. It is particularly important to do your mouth care regularly while your mouth is sore. Your nurse will show you how to care for your mouth during this time. This may include using a soft toothbrush and mild toothpaste. Avoid commercial mouthwashes, like the ones you can buy at the supermarket. These are often too strong, or they may contain alcohol which will hurt your mouth.

Bowel changes

Chemotherapy and radiotherapy can cause some damage to the lining of your bowel wall. This can lead to cramping, wind, abdominal swelling and diarrhoea. Be sure to tell the nurses and doctors if you experience any of these symptoms. If you develop diarrhoea, a specimen will be required from you to ensure that the diarrhoea is not the result of an infection. After this you will be given some medication to help stop the diarrhoea and/or the discomfort you may be feeling.

It is also important to tell the nurse or doctor if you are constipated or if you are feeling any discomfort or tenderness around your bottom (anus) when you are trying to move your bowels. You may need a gentle laxative to help soften your stool.

Hair loss

For most of us, the thought of losing our hair is very frightening. Hair loss is unfortunately a very common side effect of chemotherapy and some forms of radiotherapy. It is, however, usually only temporary. The hair starts to fall out within a couple of weeks of treatment and tends to grow back three to six months later. In the meantime there are lots of things that you can do to make yourself feel more comfortable.

Avoiding the use of heat or chemicals and only using a soft hairbrush and a mild baby shampoo can help reduce the itchiness and scalp tenderness which can occur while you are losing your hair. When drying your hair, pat it gently rather than rubbing it with a towel. Some people find it more comfortable to simply have a short hair cut when they notice that their hair is starting to fall out.

You need to avoid direct sunlight on your exposed head (wear a hat) because chemotherapy and radiotherapy makes your skin even more vulnerable to the damaging effects of the sun (i.e. sunburn and skin cancers). Remember that without your hair, your head can get quite cold, so a beanie might be useful, especially if you are in an air-conditioned environment like a hospital. Hair can also be lost from your eyebrows, eyelashes, arms and legs.

'Look Good, Feel Better' is a free community service that runs programs on how to manage the appearance-related side effects of cancer treatments. The beauty therapists who run these programs give useful advice and demonstrations on how to manage hair loss including the use of hats, wigs, scarves or turbans. You might like to find out more or register for a workshop, call 0800 865 432.

Fatigue

Most people experience some degree of tiredness in the days and weeks following chemotherapy and radiotherapy. 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.

Side effects of cortico-steroids

The types of side effects seen with cortico-steroids 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.

Cortico-steroids 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 cortico-steroids 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 cortico-steroids are temporary and should pass once you finish taking them. Long-term use of cortico-steroids 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. These effects are not common however as most people with ALL do not require prolonged steroid therapy.

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

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.

There are some options for preserving your fertility, if necessary, while you are having treatment. These are described below.

Protecting your fertility - Men

Sperm banking is a relatively simple procedure whereby the man donates semen, which is then stored at a very low temperature (cryopreserved), with the intention of using it to achieve a pregnancy in the future. You should discuss sperm banking with your doctor before starting any treatment that might impact on your fertility. In some cases, however, people are not suitable for sperm banking when they are first diagnosed because they are too unwell and therefore unable to produce the sperm in sufficient quantity or quality.

If possible, semen should be donated on more than one occasion. It is important to realise that there are many factors that can affect the quality and quantity of sperm collected in a semen donation and its viability after it is thawed out. There is no guarantee that you and your partner will be able to achieve a pregnancy and healthy newborn in the future. You should raise any concerns you have with your doctor who can best advise you on your fertility options.

The use of donor sperm might be another option for you and your partner. The sperm is donated from another male to achieve a pregnancy.

Protecting your fertility - Women

There are several approaches that may be used to protect a woman's fertility. These are outlined below.

Embryo storage - this involves collecting your eggs, usually after taking drugs to stimulate your ovaries to produce a number of eggs, so that more than one egg can be collected. This process takes at least several weeks and this can be a problem if your treatment needs to start immediately. Once the eggs are collected they are then fertilised with your partner's sperm and stored to be used at a later date. Your unfertilised eggs can also be collected and stored in a similar manner (egg storage).

Ovarian tissue storage - this is still a fairly new approach to protecting your fertility and to date there is very little experience with this technique in New Zealand. It involves the removal and storage at a very low temperature of some ovarian tissue (cryopreservation). It is hoped that at a later date the eggs contained in this tissue can be matured, fertilised and used to achieve a pregnancy.

To date, these first two approaches have unfortunately shown little success in cancer patients.

The use of donor eggs might be another option for you and your partner. These eggs could be fertilised using your partner's sperm and used in an attempt to achieve a pregnancy in the future.

It is important to understand that these methods are still quite experimental and for many reasons achieving a pregnancy and subsequently a baby is not guaranteed by using any of them. In addition, some are time consuming and costly while others may simply not be acceptable to you or your partner.

Because of the need to start treatment without delay and the problems associated with the leukaemia itself, it is often not possible to collect eggs or ovarian tissue prior to the first cycle of chemotherapy.

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.

Menstruation

Cancer treatment can also affect your periods; you may find your periods stop or become irregular. You may be prescribed a birth control pill to stop your periods. This prevents heavy bleeding and blood loss when your platelets are low. If you are having chemotherapy, it is best to use pads instead of tampons if you are menstruating as this will reduce the risk of infections. Always let your doctor know if you are having your period.

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.

Supportive care

Supportive care plays an important role in the treatment of many people living with leukaemia. This involves making every effort to improve your quality of life, by relieving any symptoms you might have and by preventing and treating any side effects that arise from your disease or treatment. Blood transfusions, antibiotics, and for some people, complementary therapies, are all important elements of supportive care.

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 ALL.

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.

Making treatment decisions

Many people feel overwhelmed when they are diagnosed with ALL. 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.

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 or some paper and a pen by your bedside as many questions are thought of in the early hours of the morning.

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, for example 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 overprotective 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.

Antibodies
Naturally produced substances in the blood, made by white blood cells called B-lymphocytes or B-cells. Antibodies target antigens on foreign or abnormal substances such as bacteria, viruses and some cancer cells and cause their destruction.

Antiemetic
A drug which prevents or reduces feelings of sickness (nausea) and vomiting (emesis).

Antigen
A substance, usually on the surface of a foreign body such as a virus or bacteria that stimulates the cells of the body's immune system to react against it by producing antibodies.

Blast cells
Immature blood cells normally found in the bone marrow.

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 many flat or big bones of the body. The bone marrow contains stem cells from which all blood cells are made.

Blood count
A routine blood test that measures the number and type of cells circulating in the blood.

Cancer
A disease characterised by uncontrolled production, accumulation and maturation of cells; often called malignant disease or neoplasm. Cancer cells grow and multiply, eventually causing a mass of cancer cells known as a tumour.

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

Central venous catheter (CVC)
A line tube passed through the large veins of the arm, neck, chest or groin and into the central blood circulation. It may be used for taking samples of blood, giving intravenous fluids, blood, chemotherapy and other drugs without the need for repeated needles.

Chemotherapy
Treatment using anti-cancer drugs. Single drugs or combinations of drugs may be used to kill and prevent the growth of cancer cells. Although aimed at cancer cells, chemotherapy can also affect rapidly dividing normal cells and therefore causes some common side-effects including hair loss, nausea and vomiting, and mucositis. The side-effects of chemotherapy are usually temporary and reversible.

Complete remission
Anti-cancer treatment has been successful and so much of the disease has been destroyed that it can no longer be detected using current technology. In people with leukaemia this means that proportion of blast cells in the marrow has been reduced to less than five per cent. There are no blast cells present in the circulating blood and the blood count has returned to normal.

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

Cytogenetic tests
The study of the structure of chromosomes. Cytogenetic tests are carried out on samples of blood and bone marrow to detect chromosomal abnormalities associated with disease. This information helps in the diagnosis and selection of the most appropriate treatment.

Disease progression
This means that the disease is getting worse despite treatment.

Echocardiogram
A special ultrasound scan of the heart.

Electrocardiogram (ECG)
Electrical trace of the heart.

Growth factors
A complex family of proteins produced by the body to control the production 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 cell transplantation. For example G-CSF (granulocyte colony stimulating factor).

Haemopoiesis (also called haematopoiesis)
The formation of blood cells.

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

Hickman catheter
A type of central venous catheter sometimes used for patients undergoing intensive treatment including bone marrow or peripheral blood cell transplantation. It may have a single, double or triple tube or lumen.

High-dose therapy
The use of higher than normal doses of chemotherapy to kill off resistant and left over cancer cells.

Immune suppression
The use of drugs to reduce the function of the immune system.

Immunocompromised
When the function of the immune system is reduced.

Immunophenotyping
Specialised laboratory test used to detect markers on the surface of cells. These markers identify the origin of the cell.

Inversion
Where parts of a chromosome turn upside down or when two parts of a chromosome reverse their positions.

Leukaemia
Cancer of the blood and bone marrow characterised by the widespread, uncontrolled production of large numbers of abnormal and / or immature blood cells. These cells crowd the bone marrow and spill out into the bloodstream.

Leukaemic blasts
Abnormal blast cells which multiple 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.

Localised disease
Disease that is confined to a small area or areas.

Lymph nodes or glands
Structures found throughout the body, for example in the neck, groin, armpit, chest and abdomen, which contain both mature and immature lymphocytes. There are millions of very small lymph glands in all organs of the body.

Lymphoid
Term used to describe a pathway of maturation of blood cells in the bone marrow. White blood cells (B-lymphocytes and T-lymphocytes) are derived from the lymphoid stem cell line.

Lymphocytes
Specialised white 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.

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

Mucositis
An inflammation of the lining of the mouth, throat or gut.

Myeloid
Term used to describe a pathway of maturation of blood cells in the bone marrow. Red blood cells, white blood cells (neutrophils, eosinophils, basophils and monocytes) and platelets are derived from the myeloid stem cell line.

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.

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

Partial remission
The tumour shrinks to less than half its original size after treatment. In people with leukaemia, this means that the proportion of blast cells in the marrow has been reduced, following treatment but not necessarily below five per cent. There are still some leukaemic cells present.

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

PICC line
Peripherally inserted central venous catheter (see central venous catheter). It is inserted in the middle of the forearm. PICCs are sometimes used for people having chemotherapy.

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.

Relapse
The return of the original disease.

Resistant or refractory disease
This means that the disease is not responding to treatment.

Spleen
An organ that accumulates lymphocytes, acts as a reservoir for red blood cells for emergencies, and destroys red blood cells, white blood cells and platelets at the end of their lifespan. The spleen is found high in the abdomen on the left-hand side. It is often enlarged in diseases of the blood or bone marrow.

Splenomegaly
Enlargement of the spleen.

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 (blood) stem cells have the ability to grow and produce all the different blood cells including red cells, white cells and platelets.

Stem cell transplant (haemopoeitic or blood stem cell transplant)
General name given to bone marrow and peripheral blood stem cell transplants. These transplants are used to support the use of high-dose chemotherapy and/or radiotherapy in the treatment of a wide range of cancers including leukaemia, lymphoma, myeloma and other diseases.

Standard therapy
The most effective and safest therapy currently being used.

T-lymphocyte
A type of white cell involved in controlling immune reactions.

Translocation
When a chromosome or part of a chromosome migrates onto another chromosome.

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 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.

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

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

- ☐ Dictionary of Terms
- ☐ Haematology Patient Diary
- ☐ Clinical Trials
- ☐ Autologous Stem Cell Transplants
- ☐ Allogeneic Stem Cell Transplants
- ☐ Myeloproliferative Disorders
- ☐ Myelodysplastic Syndromes
- ☐ Myeloma
- ☐ Acute Lymphoblastic Leukaemia in Adults
- ☐ Acute Lymphoblastic Leukaemia in Children
- ☐ Acute Myeloid Leukaemia
- ☐ Chronic Lymphocytic Leukaemia
- ☐ Chronic Myeloid Leukaemia
- ☐ Hodgkin Lymphoma
- ☐ Non-Hodgkin Lymphoma
- ☐ My Guide to Blood Cancer - for adolescents and young adults

Or information on:

- ☐ Leukaemia & Blood Cancer New Zealand's Support Services
- ☐ How to make a bequest to Leukaemia & Blood Cancer New Zealand

Newsletters:

- ☐ LifeBlood
- ☐ Lymphoma Today
- ☐ Leukaemia Today
- ☐ Myeloma Today

Name: _____

Address: _____

Postcode: _____ Phone: _____

Email: _____

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

Leukaemia & Blood Cancer New Zealand will record your details to facilitate services and keep you informed about leukaemia and related blood disorders. We value your privacy and take all the necessary steps to protect it. You can access, change or delete this information by contacting us at info@leukamia.org.nz

Notes

Notes

Acute Lymphoblastic Leukaemia in Adults

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?

☐ Yes ☐ No

Comments

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?

6. What did you like least about this booklet?

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

Contact details of Haematology Centres throughout NZ

Centre	Address	Phone
Whangarei Hospital	Hospital Road, Whangarei	(09) 430 4100
North Shore Hospital	Shakespeare Road, Takapuna	(09) 486 1491
Auckland Hospital	Park Road, Grafton	(09) 379 7440
Starship Hospital	Park Road, Grafton	(09) 379 7440
Middlemore Hospital	Hospital Road, Otahuhu	(09) 276 0000
Waikato Hospital	Pembroke Street, Hamilton	(09) 839 8899
Thames Hospital	Mackay Street, Thames	(07) 868 6550
Tauranga Hospital	Cameron Road, Tauranga	(07) 579 8000
Hastings Hospital	Omahu Road, Hastings	(06) 878 8109
Rotorua Hospital	Pukeroa Street, Rotorua	(07) 348 1199
Whakatane Hospital	Stewart Street, Whakatane	(07) 306 0999
Palmerston North Hospital	Ruahine Street, Palmerston North	(06) 356 9169
Wellington Hospital	Riddiford Street, Newtown	(04) 385 5999
Christchurch Hospital	Riccarton Avenue, Christchurch	(03) 364 0640
Dunedin Hospital	Great King Street, Dunedin	(03) 474 0999
Invercargill Hospital	Kew Road, Invercargill	(03) 218 1949

leukaemia & blood cancer new zealand

our mission is to care, our vision is to cure

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