

# Acute Lymphoblastic Leukaemia (ALL) in Children

A guide for parents, families & whānau



our mission is to care, our vision is to cure

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



Leukaemia & Blood Cancer New Zealand is grateful to Farmers Trading Company for sponsoring this booklet

## Introduction

This booklet has been written to help parents, families or whanau understand more about acute lymphoblastic leukaemia (ALL) in children.

If your child or a child that you care for has been diagnosed with leukaemia, you may be feeling anxious or a little overwhelmed. This is normal. Perhaps they have already started treatment or you are discussing different treatment options with doctors and 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 child's 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 most useful 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 separate '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 child's treatment centre.

We use the word 'family' throughout this booklet to mean those who are closest to the child. This may include parents, brothers and sisters, grandparents, other family members and friends.

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

Leukaemia & Blood Cancer New Zealand acknowledges the support of the Leukaemia Foundation of Australia for granting us permission to use material within this booklet.

Leukaemia & Blood Cancer New Zealand also gratefully acknowledges Dr Nyree Cole (Starship Children's Hospital), Dr Siobhan Cross (Christchurch Hospital) and members of the multi-disciplinary team at Starship Children's 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](mailto:info@leukaemia.org.nz) or by visiting [www.leukaemia.org.nz](http://www.leukaemia.org.nz).

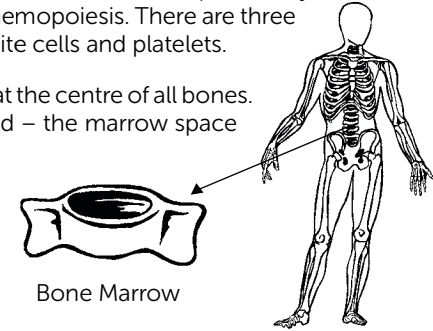
We welcome visitors to our offices in Auckland, Wellington and Christchurch. Please phone for an appointment.

## 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.

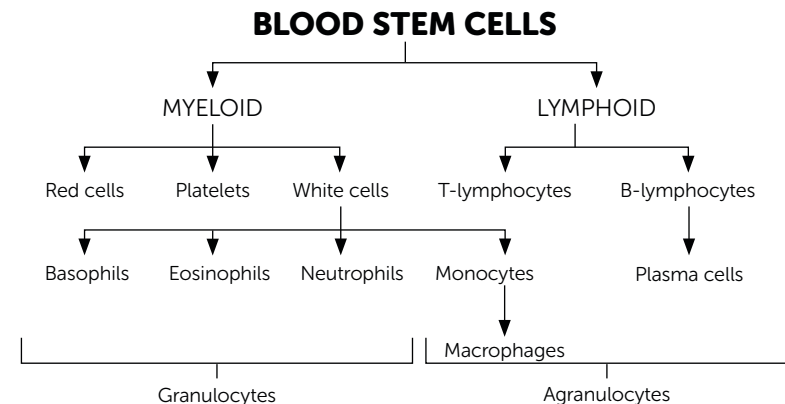
As an infant, haemopoiesis takes place at the centre of all bones. As an adult, fewer new cells are needed – the marrow space in the arms and legs is replaced by fat, and active marrow is limited to the hips, ribs and breastbone (sternum). Some of you may have had a bone marrow biopsy taken from the bone at the back of your hip (the iliac crest) or the breastbone.



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

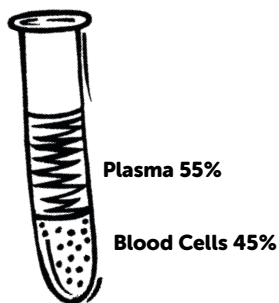
**Myeloid** ('my-loid') stem cells develop into red cells, white cells (neutrophils, eosinophils, basophils and monocytes) and platelets.

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



## Blood

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



## Blood cells

Note: The normal blood counts provided in this section of the booklet may differ slightly from the ones used at your child's treatment centre. You can ask for a copy of your child's blood results, which will include the normal values for each blood type.

### Red cells and haemoglobin

Red cells contain haemoglobin (Hb), which gives the blood its red colour and 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.

Normal ranges of haemoglobin for children:

	1 month	1 year	3 years	5 years	9 years	16 years
<b>Haemoglobin (g/L)</b>	102-130	104-132	107-136	110-139	113-143	115-165 (F) 130-180 (M)

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 your child is anaemic they may feel run down and weak. They may be pale and short of breath or they may tire easily. In this situation a red cell transfusion may be given to restore the red cell numbers and therefore the haemoglobin to more normal levels. Red cell transfusions are given depending on an individual child's symptoms and phase of treatment, not just to treat the haemoglobin level on the blood test.

## 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

When your child's white cell count drops below normal they are at risk of infection.

Normal white cell count for children:

	1 month	1 year	3 years	5 years	9 years	16 years
<b>White cells (<math>\times 10^9/L</math>)</b>	6.4-12.1	5.4-13.6	4.9-12.8	4.7-12.3	4.7-12.2	3.5-11

Neutropenia is the term given to describe a lower than normal neutrophil count. If your child is neutropaenic (neutrophil count of less than  $1.0 \times 10^9/L$ ) they are considered to be at risk of developing frequent and sometimes severe bacterial or fungal infections.

Normal neutrophil count for children:

	1 month	1 year	3 years	5 years	9 years	16 years
<b>Neutrophils (<math>\times 10^9/L</math>)</b>	0.8-4.9	1.1-6.0	1.7-6.7	1.8-7.7	1.8-7.6	1.7-7.0

## 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 injury, stick together and form a plug to help stop the bleeding.

Normal platelet count for children:

	1 month	1 year	3 years	5 years	9 years	16 years
<b>Platelets (x 10<sup>9</sup>/L)</b>	270-645	205-553	214-483	205-457	187-415	150-450

Thrombocytopenia is the term used to describe a reduction in the platelet count to below normal. If your child's platelet count is low, they 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 platelet level falls below  $10 \times 10^9/L$ .

## Growth factors and cytokines

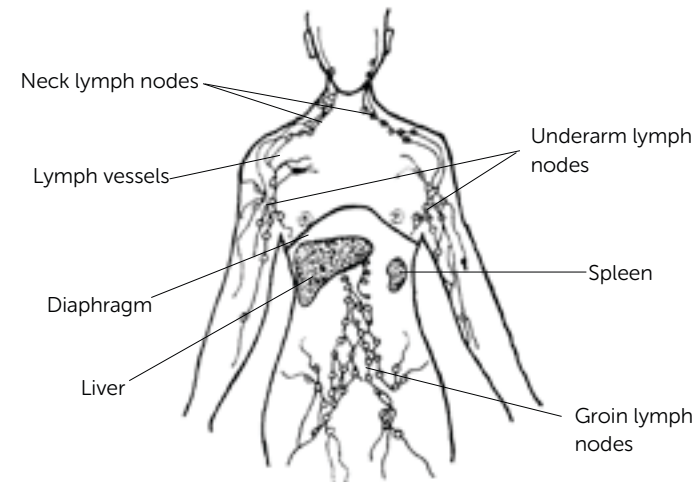
All normal blood cells have a limited survival in the circulation and need to be replaced on a continual basis. This means that the bone marrow remains a very active tissue throughout your life. Natural chemicals in your blood called growth factors or cytokines control the process of blood cell formation. 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 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 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. Acute leukaemia affects very immature blood cells, preventing them from maturing properly.

In chronic leukaemia there is an accumulation of more mature but abnormal white cells. Chronic leukaemia can occur at any age but is more common in older adults. It 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 conditions these cells grow and mature into specialised white cells called B-lymphocytes (B-cells) and T-lymphocytes (T-cells). In ALL, they undergo a malignant (cancerous) change. This means that 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, children with ALL become more susceptible to anaemia, recurrent infections and to bruising and bleeding easily.

Excess numbers of 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, central nervous system (brain and spinal cord) and testes. ALL may present as lymphoma (cancer of the lymph nodes) if the majority of the leukaemic cells are first found in the lymph nodes. Despite the different name (acute lymphoblastic lymphoma) this is basically this same disease and is treated in the same way.

Improvements in the diagnosis and treatment of children with ALL means that, the majority of children treated for ALL today will achieve a remission from their disease and most will be cured.

## How common is ALL and who gets it?

Each year in New Zealand around 40 children aged between 0-14 years are diagnosed with leukaemia. Of these ALL is the most common type with approximately 33 children each year being diagnosed.

Children can also develop other types of leukaemia such as acute myeloid leukaemia (AML), chronic myeloid leukaemia (CML) and other types of blood cancers like lymphomas.

## What causes ALL?

When a child is diagnosed with ALL, parents naturally want to know what has caused this disease. No one knows exactly what causes ALL, but it is likely that there are a number of factors, rather than any single factor involved. Research is ongoing into possible causes and a number of environmental factors continue to be investigated. To date, none have been proven to cause ALL in children.

It is important to realise that you, as a parent, have not caused your child's disease. Like many cancers, ALL is thought to result from a series of changes in special proteins called genes, which normally control the growth and division of cells. The reasons for these changes remain unclear. There are certain factors that may put some children at a higher risk of this type of genetic damage and therefore the development of ALL. These are called risk factors and they are described below.

### **Ionising radiation**

Children exposed to large doses of ionising radiation (a type of energy emitted from x-rays and radioactive materials) before they were born or in the early years of life may be more at risk of developing leukaemias like ALL. These include the survivors of the nuclear bombs in Japan at the end of World War II. It is unlikely that any children born in New Zealand are exposed to high enough levels of ionising radiation to cause childhood ALL.

### **Chemicals**

Exposure to high levels of benzene and other industrial solvents over a long period of time may increase the risk of some blood disorders like leukaemia. Children in New Zealand are unlikely to be exposed to high enough levels of these chemicals to cause ALL.

### **Infections**

There is some evidence to suggest that viral infections may play a role in the development of ALL in some children. No specific virus has been implicated. It is thought that delayed exposure to common childhood infections or an abnormal response by the child's immune system to these infections may be involved. This is supported by the higher incidence of ALL reported in

particular geographic or demographic areas. ALL is not contagious, that is, a child cannot 'catch' ALL by being in contact with someone who has it.

### **Electro-magnetic radiation**

In recent years there has been a great deal of controversy about the health effects of living very close to high-voltage power lines and other sources of electro-magnetic radiation such as mobile phones, mobile phone base towers and electrical equipment in our homes. The results of several large international studies have provided no clear evidence to support a link between childhood ALL and exposure to acceptable levels of electro-magnetic radiation in our environment.

### **Genetic factors**

Although childhood ALL is not inherited, genetic factors may play a role in its development. Children with certain congenital disorders like Down's syndrome are at an increased risk of developing ALL. Some children appear to be born with genetic changes that increase the risk of developing childhood leukaemia.

## What are the symptoms of ALL?

Because ALL develops quickly, children 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, children 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 children have frequent or severe nosebleeds or bleeding gums. Red or purple flat pinhead sized spots may appear on the skin, especially on the legs. These are called petechiae ('pe-tee-key-eye') and they are caused by tiny bleeds under the skin.



## Frequent or repeated infections

Children with ALL don't have enough normal white blood cells so they are more likely to develop frequent or repeated infections. These may present as minor skin infections, a sore throat, and sore mouth or slow healing of minor cuts and grazes. They may also develop chest infections (coughing), urinary tract infections (frequent passing of urine with a sensation of burning) and fevers. The leukaemia itself can be the cause of low grade fever, in the absence of an infection.

## Bone pain

Pain in the bones and joints is common and results from the marrow being literally crowded with leukaemic cells. Occasionally there may be deposits of leukemic cells in bone itself and this can cause localised pain.

Other symptoms of ALL may include swollen lymph nodes (glands), chest pain and abdominal discomfort due to a swollen spleen or liver.

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

## 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) also known as a complete blood count (CBC). This involves taking a sample of your child's blood, usually from a vein in their 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 children with ALL have a low red cell count, a low haemoglobin level, and a low platelet count. Most children have a high white cell count and almost all children will have abnormal leukaemic blast cells in their bloodstream. While the presence of leukaemic blast cells in your child's bloodstream suggests that they may have leukaemia, the diagnosis will need to be confirmed by examining their bone marrow cells.

Your child's blood count will be checked regularly both during and after treatment to see how well they are progressing and how well their disease is responding to treatment.

## Bone marrow examination

If the result of your child's blood count is abnormal and suggestive of ALL, a bone marrow examination will be needed to confirm the diagnosis, and to decide on the best possible treatment for your child. This involves taking small samples of your child's bone marrow, usually from the back of the hipbone, and sending it to the laboratory for examination.

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 (usually less than 5 per cent) of normal developing blood cells, known as blast cells. This proportion can increase to between 20% and 95% in children with ALL.

The bone marrow examination will be done in the hospital. Most children receive a short general anaesthetic for this procedure. In some centres, older children and adolescents may have a local anaesthetic, some painkillers and sedation. The doctors and nurses at the hospital will discuss with you the most appropriate choice for your child. Samples of bone marrow are collected using a long thin needle 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'. In some instances, 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'.

After the procedure is finished a small dressing or plaster is placed over the needle site. This can usually be removed the next day. Your child may have some mild bruising or discomfort, which is usually managed effectively with paracetamol. More serious complications such as bleeding or infection are very rare.

During treatment your child will need repeat bone marrow examinations to assess how well the disease is responding.

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 your child has, 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.

The Ph chromosome occurs in < 5% of children with ALL but it is the most common chromosomal abnormality seen in adult ALL.

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 child's disease has responded to treatment.

We now know that there is usually a strong relationship between the number of leukaemic cells leftover in a child's body following treatment, and their risk of relapse in the future. Using newer technologies, it is now possible to measure this leftover or minimal residual disease (MRD), normally not visible

under the microscope. Measuring MRD has become a standard way of testing a child's response to initial treatment, their future risk of relapse and therefore, the most appropriate treatment protocol for their particular circumstances. MRD testing can also be repeated at various points along the way to assess how well your child is progressing, and responding to a chosen treatment.

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

## Other tests

Other tests provide information on your child's general health and how well their 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 child's blood is clotting properly.

Your child may also have a procedure called a lumbar puncture. During which a small sample of the cerebro-spinal fluid (CSF) that surrounds the brain and spinal cord is taken via a needle in the lower back. This fluid is tested in the laboratory to check for the presence of leukaemia 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 child's general health. The results may be important in selecting the best treatment option for them. The results can also be compared with later results to assess how well your child is progressing.

## Which type of ALL does my child 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 page 15), 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 16)

## Pre-B-cell ALL

In around 80% of cases, childhood ALL arises in B-lymphocytes (B-cells) 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. In the majority of precursor B-cell ALL (around 80%), the common ALL antigen known as cALLa, or CD10 is expressed on the surface of the leukaemic cells. Precursor-B-cell ALL can be further classified into early pre-B-cell, pre-B, or transitional pre-B-cell ALL, depending on antigens expressed on the leukaemic cell surfaces.

## B-cell ALL

B-cell ALL arises in more mature developing lymphocytes. This type of ALL is less common, accounting for around 5% of all cases. Here, leukaemic cells tend to spread to areas outside the blood and bone marrow and collections of leukaemic lymphoblasts may be found in the abdomen, head, and neck regions. Involvement of the central nervous system is common.

B-cell ALL is biologically very similar to another disease called Burkitt's lymphoma, a rare, aggressive type of lymphoma. Children diagnosed with B-cell ALL are generally treated with similar drugs to those used to treat this lymphoma.

## T-cell ALL

In around 15% of cases, ALL arises in developing T-cells in the thymus gland in the chest. Precursor-T-cell ALL can be further classified as early-, mid- or late- thymocyte-T-cell ALL, depending on the maturity of the affected cell. Children with T-cell ALL often have a high white blood cell count and involvement of the central nervous system at diagnosis. In around 50% of cases, the thymus gland is enlarged and visible on x-rays in the centre of the chest (mediastinal mass).

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

## Prognosis

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

Your child's doctor is the best person to give you an accurate prognosis regarding your child's leukaemia as he or she has all the necessary information to make this assessment. An accurate prognosis cannot usually be given until the cytogenetic test results are known and the speed of the response assessed. The prognosis may be altered depending on the result of this test.

While the outlook for most children with ALL is very good, certain factors (known as prognostic factors) give some children a better chance of being cured of their disease with treatment than others. The most important of these factors is how well your child's disease responds to initial treatment, or in other words, how quickly they achieve a remission and how much disease is left over in the body after this initial treatment.

Other related factors include the age and sex of your child, the exact type of disease they have, their white cell count at diagnosis and whether or not the leukaemia had spread to the central nervous system (CNS) at the time of diagnosis. The genetic make-up of the leukaemic cells is another important factor in predicting prognosis and the likelihood of cure in ALL. For example, leukaemia expressing the abnormal Philadelphia chromosome has been associated with a poorer prognosis using standard therapy.

Taking these and other factors into consideration, children are categorised as having low, standard or high-risk ALL. This ensures that the most appropriate and effective 'risk-based' therapy can be chosen for every child. For example, intensive therapy may be more beneficial than standard therapy for a child who belongs to the high-risk group. Intensive therapy will help to reduce the child's risk of future relapse and therefore increase their overall chances of survival. It is important to realise that although almost all children treated for ALL will achieve a remission, a small proportion will experience a relapse over time.

## Commonly used prognostic terms

The following terms may be used to describe how well the ALL 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. With treatment, the majority of children with ALL can be cured of their disease.

**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%. There are no blast cells present in the circulating blood and the blood count has returned to normal and no cytogenetic abnormalities can be detected.

Almost all children with ALL will achieve a remission. The length of time that a remission lasts may vary from child to child, and the leukaemia may well reappear (relapse) over time.

**Resistant/refractory disease** - This means that the leukaemia is not responding to treatment.

**Relapse** - The leukaemia has reappeared. This can be in the bone marrow (most common site), the sanctuary sites, for example the CNS or testis, and occasionally other sites such as the bone or lymph glands.

## How is ALL treated?

ALL usually progresses quite quickly so treatment needs to begin as soon as it is diagnosed. Although the diagnosis may be straightforward and made rapidly, occasionally it is more complicated. Under these circumstances it is obviously important to take time to be sure the diagnosis is absolutely certain.

Children diagnosed with ALL need to be treated in a specialist paediatric cancer centre under the care of a specialist doctor called a paediatric haematologist/oncologist. A paediatric haematologist/oncologist is a doctor who specialises in the care of children and adolescents with cancer and diseases of the blood, bone marrow and immune system. Your child's treating doctor and other members of the treatment team will keep your general practitioner (GP) and/or local paediatrician informed about your child's condition so that their care can be shared between the specialist centre and your local hospital/GP service further down the track.



Children in New Zealand who are diagnosed with ALL usually follow established protocols or plans of treatment as part of international research studies (clinical trials) into improving the way this disease is treated. These protocols can vary between children and the particular institution at which a child is being treated. The treatments given as part of each protocol are standardised. This means that hundreds of children around the world participating in the same trial and allocated to the same protocol (as your child) will receive the same treatment. In this way important information can be collected which will continue to improve the way in which children with ALL are treated in the future.

The type of protocol your child is allocated to will depend on the 'risk group' to which they belong. The risk group to which they belong will be defined based on a number of clinical and laboratory factors, both at diagnosis and during treatment, that predict the outcomes of particular treatment approaches. Your child's progress and response to treatment is closely monitored throughout all phases of their treatment. Sometimes adjustments need to be made to your child's protocol depending on how well they are responding to treatment.

The treatment of ALL can last from two to three years or longer depending on your child's particular circumstances, the treatment protocol they are following and how well they are responding to treatment. It is important to realise that whatever protocol your child follows, it is the best treatment available at this time.

## Clinical trials

Clinical trials (also called research studies) test new treatments or existing treatments given in new ways to see if they work better. The information gathered from clinical trials has contributed to the high cure rates and survival rates for children with ALL. These trials continue to be important because they provide vital information about how to further improve treatment by achieving better results with fewer side effects. In addition, clinical trials often give people access to new therapies not yet funded by governments.

As parents you will need to give your informed consent (see below) for your child's participation in a clinical trial. Your child's doctor will discuss with you the best treatment options for your child. He or she will also provide you with information that will help you to understand the reasons for a particular clinical trial, the benefits and risks of the trial and what it involves for your child and your family. You need to have this information before you can give your informed consent.

There is a separate booklet about clinical trials available from Leukaemia & Blood Cancer New Zealand.

## Informed consent

Giving your informed consent means that you, as the child's parent or guardian, understand and accept the risks and benefits of a proposed procedure or treatment for your child. It means that you are happy that you have adequate information to make such a decision.

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

If you have any doubts or questions regarding any proposed procedure or treatment do not hesitate to ask for more information from the doctor.

## 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 child's age and general health, and the treatment protocol that is being followed.

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 given in several phases with time for recovery at the end of each phase. This is to allow your child's body (the bone marrow in particular) time to recover from the side effects of chemotherapy.

Chemotherapy is given in many different ways in the treatment of ALL. Some drugs are given in tablet or liquid form (orally); others may be injected into a vein (intravenously or IV), into a muscle (intramuscularly or IM), and under the skin (subcutaneously or SC). Chemotherapy is also given intrathecally (into the spinal fluid or IT), through a lumbar puncture, to either treat or prevent the spread of leukaemic cells into central nervous system (CNS).

Intravenous 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 child's 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.

After their initial treatment your child may be able to receive a majority of the rest of their treatment in the outpatient department of the hospital or clinic, or at home. However, depending on the type of chemotherapy being given or your child's general health, they may need to be admitted to hospital.

## Cortico-steroid therapy

Cortico-steroids (also known as '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, prednisolone 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 central nervous system (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 child's treatment. This usually involves injections of methotrexate and/or other chemotherapy drugs directly into the spinal fluid (called an 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.

### Testicular radiotherapy

The testes in boys can also act as a 'sanctuary site' for leukaemic cells but unless disease is found here at diagnosis, no additional treatment is required. Your child's doctor will decide on the most appropriate treatment in the event of testicular disease. This may or may not include radiotherapy. High dose chemotherapy may also be used.

### 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 your child is diagnosed, they will need to begin an intensive course of treatment to bring about (induce) a remission. The goal of remission induction therapy is to destroy any detectable leukaemic cells in your child's blood and bone marrow and allow their bone marrow to function normally again. Your child will need to be admitted to hospital for part or all of this first phase of treatment.

Commonly used chemotherapy drugs in this phase of treatment include: vincristine, daunorubicin, asparaginase and cortico-steroids. Not all children will need all of these drugs. CNS therapy also begins at this stage.

While your child is undergoing induction therapy they may also be given a drug called allopurinol. This 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 who are at high risk of this complication (such as those who have a very high leukaemia cell count) a new drug called rasburicase may be used to protect the kidneys.

Almost all children with ALL will achieve a remission following induction therapy. However, in a small number of cases, the disease will not respond to treatment as expected. In this situation, the child 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 child's disease more effectively.

### Consolidation

Soon after remission induction therapy finishes, more treatment is required to help destroy any leftover disease in your child's body. 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 therapy or intensification. The consolidation protocol chosen for your child will depend on their estimated risk of relapse in the future, in other words the 'risk group' to which they belong.

## Maintenance

Maintenance therapy is designed to help keep your child's disease in remission and prevent it from reappearing (relapsing) in the future. Common maintenance protocols involve chemotherapy tablets taken daily and in some protocols also injections of chemotherapy with courses of cortico-steroids given monthly. In addition, intrathecal injections of chemotherapy may be given periodically to prevent disease relapsing in the CNS.

This phase of treatment will continue until the treatment is completed. This is a total treatment time of just over two years for girls, and just over three years for boys. During this time, your child will be treated as an outpatient.

After initial induction treatment children are encouraged to take part in their usual daily activities including attending school or day care, as they are able. Your doctor will advise you when it is safe for your child to return to these activities and when it is safe to continue immunisations, which are usually delayed until 6–9 months after your child has finished treatment. If your child has a stem cell transplant, immunisations may be delayed for 6-12 months afterwards.

While your child is receiving maintenance therapy, they will be examined regularly by the doctor who will do a full physical examination and check their blood counts. During this time the doctor will make an assessment of how well your child is progressing, and adjust their treatment as necessary.

### Stem cell transplant

For a small number of children, the chance of curing ALL with chemotherapy alone may be low. If these children have a sibling who is of a similar tissue type, or if a suitable unrelated donor can be found on the international registries, the doctors may recommend a haemopoietic stem cell transplant (previously called a bone marrow transplant. Now, the source of cells may be from bone marrow, blood or umbilical cord blood). This relies on very high doses of chemotherapy and/or radiotherapy to treat your child's disease more effectively.

Due to the complex side effects associated with this form of treatment and the success of current protocols used to treat ALL, a stem cell transplant is usually only offered in selected cases where the doctor feels that it will benefit a particular child. For example, in the case of very high-risk disease, relapsed disease, or disease which is proving resistant to conventional treatment.

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

## Relapsed disease

Finding out that your child's 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 child's age and the genetic make-up of the relapsed leukaemic cells. Similar drugs to those used to initially treat leukaemia, different drugs, and in some cases, high dose chemotherapy and a stem cell transplant may be used to treat relapsed disease.

Late relapse (relapse that occurs years later) is usually more responsive to further treatment than relapse that occurs soon after a remission has been achieved. Clinical trials are continuing to determine the best way to treat relapsed ALL to achieve the best outcome for all children.

## Palliative care

If a decision is made not to continue with anti-cancer treatment (chemotherapy and radiotherapy) for your child's leukaemia there are still many things that can be done to help them 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

Children react differently to treatment. The type and severity of side effects can vary from child to child, depending on the type of treatment used and how an individual child responds to it. In general, more intensive treatment is associated with more severe side effects.

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

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

- a temperature of 38°C or higher (even if it returns to normal) and/or an episode of uncontrolled shivering (a rigor)
- bleeding or bruising, for example blood in the urine, bowel motions, coughing up blood, bleeding gums or a persistent nose bleed
- prolonged nausea or vomiting that prevents them from eating or drinking or taking their normal medications
- diarrhoea, stomach cramps or severe constipation
- persistent coughing or shortness of breath or increased respiratory rate (breathing more quickly than normal)
- a new rash, reddening of the skin, itching
- a persistent headache
- a new severe pain or persistent unexplained soreness
- a cut or other injury
- persistent pain, swelling, redness or pus anywhere on their body, especially near their central line site.

It is important to know that there can be many unscheduled admissions to hospital throughout your child's treatment.

## 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 child's mouth, gut and bone marrow.

### Effects on the bone marrow

ALL prevents your child's 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 child's blood count (the number of blood cells circulating in their blood) will generally fall within a week of treatment, increasing their risk of infection and bleeding.

**White cells** - The point at which your child's white blood cell count is at its lowest is called the nadir. During this time your child will be at a higher risk of developing an infection. At this stage they will also be neutropenic, which means that their neutrophil count is low. Neutrophils are important white blood cells that help fight infection. While your child's white blood cell count is low, sensible precautions need to be taken to help prevent infection.

These include avoiding crowds, avoiding people who are unwell, 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.

Simple measures like hand washing are an effective way to reduce the risk of infections. Ask your visitors and other family members to wash their hands before having direct contact with your child. As a precaution, family members may be advised to have the flu vaccination. In general, when they are not in hospital, there is no need for your child to stop going to school or day care at any stage during treatment provided they feel up to it. They will not contract serious infections at school provided sensible precautions are taken and the benefits of maintaining social contacts outweigh any disadvantages.

The highest risk of infection is during induction and delayed intensification phases of treatment. Some centres advise not to attend school or day care during this time. It is important to note that the risk from viral infections does remain while your child is on maintenance therapy and is not always associated with low neutrophil counts. Always speak with your treatment team for advice.

Your doctor and the nurses at your child's treatment centre will advise you on how to reduce your child's risk of infection while their white cell count is low.

If your child does develop an infection they may experience a fever, which may or may not be accompanied by an episode of shivering or shaking, which is called a rigor. If your child experiences a high temperature and/or a rigor they need to be seen by a doctor immediately. Infections can be very serious and need to be treated with antibiotics as soon as possible.

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

**Platelets** - Your child's platelet count may also be affected by the disease and by the chemotherapy they are receiving. They may become thrombocytopenic (a reduction in the number of platelets circulating in the blood). When your child's platelet count is very low, they can bruise and bleed more easily. During this time it is helpful to avoid sharp objects in the mouth such as potato chips or toys that could cut your child's gums. Using a soft toothbrush also helps to protect their gums. In addition, your child should avoid any contact sports or rough play where they might get injured easily.

It is important that your child does not become constipated during this time as a hard bowel motion/stool may damage the lining of the child's bowel and cause bleeding, or infection. Your child may have a stool-softening laxative prescribed to prevent constipation during this time.

In many cases a transfusion of platelets is given to reduce the risk of bleeding until your child's platelet count recovers.

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

### Hair loss

Hair loss is a very common side effect of chemotherapy and some forms of radiotherapy. It is usually only temporary. The hair starts to fall out within a couple of weeks of treatment starting and may come and go throughout treatment. In most cases, your child's hair will grow back completely once treatment has finished. Many young children are not worried by losing their hair and are happy to wear hats, scarves or bandannas. Older children and teenagers are often more concerned about the effects of hair loss and other changes to their appearance. Girls are often encouraged to get a wig - whilst they may never wear it, having the wig may give them the confidence to participate in everyday activities, particularly those involving friends.





### Mucositis

Mucositis, inflammation of the mouth, throat or gut is a common and uncomfortable side effect of chemotherapy. Mucositis usually starts about a week after the treatment has finished and generally goes away once your child's blood count recovers, usually a couple of weeks later. During this time your child's 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 child's temperature before giving them paracetamol as this drug can 'mask' signs of infection (a raised temperature).

It is important to keep your child's mouth and teeth as clean as possible while they are having treatment especially when their mouth is sore. This can help make them feel more comfortable while also reducing their risk of infection. Different treatment centres recommend different mouth care products. The nurse will teach you and your child how to clean the mouth and teeth during this time. This may include using a recommended mouthwash and a soft toothbrush or a soft piece of gauze wrapped around a finger to clean the teeth after every meal.

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 child's mouth.

### Diarrhoea

Chemotherapy can cause damage to the lining of your child's bowel wall. This may lead to cramping, wind, bloating and/or diarrhoea. Be sure to tell the nurses and doctors if your child is experiencing any of these symptoms. If your child does develop diarrhoea, the nurse will ask for a specimen which will be tested in the laboratory, to rule out infection as the cause. Your child's bottom can become quite sore if they are having diarrhoea. Baby wipes are a good idea for cleaning their bottom; they are clean and soft and usually gentler and less abrasive than toilet paper. It may also be necessary to apply a barrier cream to your child's bottom to help protect the skin and reduce discomfort.

### Constipation

Some chemotherapy, vincristine in particular, can cause constipation. It is important to tell the nurse or doctor if your child is constipated or if they are feeling any discomfort or tenderness around their bottom (anus) when trying to move their bowels. They may need a gentle laxative to help soften the bowel motions.

Sometimes children can have diarrhoea even though they are still constipated. This is called overflow. If your child is taking laxatives and they develop diarrhoea, it is a good idea to talk to the nurses at your treating hospital before stopping the laxatives. They will be able to advise you on the steps you need to take to help restore your child's normal bowel function.

### Nausea and vomiting

Most medications used to treat ALL in children do not cause nausea and vomiting. In some cases however anti-sickness (anti-emetic) drugs are required to help prevent these symptoms. If necessary, your child will be given anti-emetics before, and for a few days after their chemotherapy treatment. Be sure to tell the nurses and doctors if the anti-emetics are not working for your child and they still feel sick. There are many types of anti-emetics that can be tried. A mild sedative may also be used to help your child relax and reduce their fears about getting sick.

Frequent severe diarrhoea and/or vomiting may cause dehydration, which can worsen your child's condition. It is important during this time, to monitor how much fluid your child is drinking and keeping down, and whether or not they are passing much urine. If your child is losing a great deal of fluid, unable to drink fluids, or if they are not passing much urine they may need to be topped up with some intravenous fluid in the hospital day treatment centre or be admitted to hospital.

### Loss of appetite

There are lots of reasons why children may not feel like eating much during treatment, especially while they are having treatment or are in hospital. Allowing your child to eat when they are hungry, which often means snacking in between meal times, and offering them nutritious snacks and drinks throughout the day can be helpful during this time.

### Fatigue

Most children will 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 them feel better during this time. Getting out into the fresh air and doing some gentle exercise is important for your child's general feeling of wellbeing and it also may help to reduce their fatigue. It is also important to allow your child to rest when they are tired. Try to plan the day in order to get a balance in between activities and rest.

### Seizures

Intrathecal (IT) therapy is rarely associated with seizures, otherwise known as fitting. If your child does experience a seizure, they will be investigated for a cause; this will include having a head scan. If the doctor feels they may be at risk of having another seizure in the future, they will prescribe special medication to help to prevent this from happening.

## Side effects of cortico-steroids

Side effects of cortico-steroids depend largely on how long they are used for and the dose given. Again, children respond differently. An increased appetite, fluid retention, weight gain and the classic 'moon-shaped' face and swollen belly are common side effects of these drugs. Many children feel hungry all the time while they are taking cortico-steroids and frequently want to eat at all times of the day and night. These side effects are usually temporary and your child's weight and eating habits should return to normal in time once they have finished treatment. In the meantime try to encourage healthy and nutritious foods limiting the amount of high-fat (chips and chocolate), high-sugar (lollies) foods they eat.

Some children find it more difficult to get to sleep at night and to stay asleep, and some night sedation may be required. Mood swings, anxiety, restlessness and nightmares are also common side effects of steroid therapy. A child's moods and behaviours can be challenging while they are receiving steroids. While accepting that some allowances need to be made, maintenance of your normal parenting strategies is important during this time. Being consistent and setting limits on your child's behaviour can help to make them feel more secure. It can also help to prevent longer-term behavioural problems, which can cause considerable stress within any family.

An increased susceptibility to infections and high blood pressure are also recognised side effects of steroid therapy.

Long-term use of steroid therapy may cause other effects such as osteoporosis (where the bones become weak and brittle) or avascular necrosis, though these effects are not common. Remember to tell your doctors and nurses about any side effects your child is having as they can usually suggest ways to help you.

## Pneumocystis prophylaxis

Almost all children with leukaemia will be prescribed a low dose antibiotic called cotrimoxazole which is used to help prevent an infection called pneumocystis carinii. This is an organism that most children have been exposed to and it can reactivate when the immune system is compromised (such as patients on chemotherapy) and cause a life-threatening pneumonia. Cotrimoxazole prophylaxis generally continues until chemotherapy is completed. There is a group of children who are intolerant of cotrimoxazole who will be prescribed other agents.

## Shared care

In many cases, particularly if you live far from the specialist centre, arrangements will be made for some of your child's care to be given in the children's ward at your local hospital. This may just be regular blood checks, or may range from blood transfusions to the administration of chemotherapy. Such arrangements are only made where all the appropriate staff and facilities are in place for such treatments to be performed safely. There is close communication between the specialist centre and shared care hospital unit to ensure that both are kept up to date with all that is happening with your child.

## Relocating to hospital for treatment

Treatment for childhood leukaemia, especially in the early stages, requires specialist care that is only available at specialised child cancer units. As a result, many patients and family members have to spend some time away from the comfort of their own home. If you need to travel a long distance to the treatment centre, accommodation may need to be arranged for your family. You may also need some accommodation outside the hospital if your child is being treated as an outpatient. Suitable accommodation can be arranged by contacting the social worker at your treatment centre. They can also assist you with any paperwork required when making claims for financial assistance.

## Follow-up

Follow-up checks continue well beyond the end of treatment to allow careful periodic assessment of your child's general health, to monitor for disease relapse and the continued growth and development of your child. These checks are important because they allow for early detection and, where necessary, early intervention if any problems arise. Most major treatment centres now have long-term follow up clinics (sometimes called late effects clinics) where specially trained health professionals assess the long-term effects of treatments on children's growth and development. They provide support to children and their families to help them cope with any difficulties that may arise.

## Long-term effects of treatment

Most children go on to enjoy long and healthy lives after being successfully treated for ALL. Sometimes, however, the treatment can affect a child's health months or even years after it has finished. These are called long-term or late effects. Your doctor will discuss any potential long-term effects of your child's treatment and the steps that can be taken to help reduce or prevent them.

The long-term effects of treatment depend on several factors, including the types of drugs and combinations of drugs used and the individual and cumulative doses used. In general, more intensive treatments, like a stem cell transplant, and treatments that involved radiation can cause more significant long-term effects.

In children, areas of the brain that control normal growth and development are immature and therefore more sensitive to the effects of some treatments. For example radiation to the central nervous system (CNS) (now only rarely used in ALL) can cause a number of long-term problems including obesity, reproductive difficulties and delayed growth. Delayed growth can be treated using growth hormone replacement therapy. CNS radiation, and other CNS treatments (intrathecal chemotherapy and some types of intravenous chemotherapy), have also been associated with learning difficulties in some children. This is more commonly seen in younger children but is infrequent with current treatments. Your child's school progress is monitored as part of their routine follow up after treatment.

## 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.

Most children who are treated for ALL will grow up and be able to have, normal, healthy babies. For others, treatment may cause a reduction in their fertility and their ability to have children in the future. This may depend on the age of child when they were treated and the type of treatment they received. The onset of puberty can also be affected and some children may require hormonal supplements to ensure normal sexual development.

In boys, sperm production may be impaired for a while following chemotherapy but production of new sperm may become normal again in the future.

In girls, chemotherapy and radiotherapy can cause varying degrees of damage to the normal functioning of the ovaries. This will depend on the age of the child and the dose of radiotherapy or chemotherapy given. In some cases this leads to menopause (change of life) earlier than expected.

### Protecting fertility - Boys

There may be some options for preserving your child's fertility. If the treatment is likely to reduce fertility, adolescent boys can be offered sperm banking. This is a relatively simple procedure whereby the adolescent boy 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. This is only available to pubescent boys; you should discuss sperm banking with your doctor before your son starts any treatment that might impact on their fertility. In some cases, your child may be unable to donate sperm at this stage, as he may be too ill to produce the sperm in sufficient quantity or quality.

### Protecting fertility - Girls

**Ovarian tissue storage** - is still a new and experimental approach to protecting female fertility. 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. This procedure may be offered to some adolescent girls (perhaps as part of a research programme) but cannot be undertaken in young children. Unless a stem cell transplant using very high dose chemotherapy and/or total body irradiation is planned, infertility in girls is very unlikely.

To date, ovarian tissue storage is one of several techniques which remain under investigation. They have not yet been proven to be successful in allowing women to bear children. Also, because of the need to start treatment without delay and the problems associated with the leukaemia itself, it is often not possible to collect ovarian tissue prior to remission induction therapy.

**Hormonal therapy** - is being studied to see if this can reduce infertility rates. Usually this involves a monthly injection of a hormone-blocker (a GnRH antagonist) to temporarily turn off the ovaries and make them less susceptible to damage by chemotherapy drugs.

It is important to realise that every effort is made to avoid treatments known to cause significant long-term problems. This needs to be balanced however, against providing the most appropriate treatment that will give a child the best chance of being cured. Research is continuing into ways to achieving the best outcomes for children with ALL while reducing the risk and impact of any long-term effects of treatment.

## Supportive care

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

### Complementary therapies

Complementary therapies are therapies which are not considered standard medical therapies. They include yoga, exercise, meditation, prayer, aromatherapy, relaxation and herbal and vitamin supplements.

Complementary therapies should only be used to 'complement' or assist with recommended medical treatment for children with ALL. They should not be used instead as an alternative to medical treatment. It is important to realise that no complementary or alternative treatment alone has proven to be effective against childhood ALL.

It is also important that you inform your doctor if your child is using any complementary therapies or alternative therapies in case they interact with chemotherapy or other treatments your child may be receiving.

### Nutrition

A healthy and nutritious diet is important in helping your child to cope with their disease and treatment. Talk to your doctor or nurse if you have any questions about your child's diet or if you are considering making any radical changes to the way they eat. You may wish to see a nutritionist or dietician who can advise you on planning a balanced and nutritious diet.

Occasionally, treatment complications result in severe weight loss and feeding using a nasogastric tube to deliver highly nutritious supplements is required. In some cases intravenous nutrition is needed for a short period.

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



## Making treatment decisions

Most parents feel overwhelmed when their child is diagnosed with leukaemia. 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, or that they are being rushed into making a decision. It is important that you feel you have enough information about your child's illness and all of the treatment options available, so that you can take part in decisions which are being made about the best way forward for your child.



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

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

Your child's treating doctor will spend time discussing with you and your family what he or she feels is the best option for your child. Feel free to ask as many questions as you need to. You should feel that you have enough information to make the decisions that are in your child's best interests. Remember, you can always request a second opinion if you feel this is necessary. However, it is important not to delay starting treatment for ALL as this disease progresses rapidly without treatment and can quickly become life-threatening. It is very useful to have a copy of the treatment roadmap with likely dates of planned admissions to try and help organise the weeks ahead.

### 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

### Parents

Parents cope with a diagnosis of childhood leukaemia in different ways and there is no right, wrong or standard reaction. Hearing that your child has been diagnosed with leukaemia is extremely distressing and can trigger a range of intense emotional responses, ranging from denial to devastation. It is not uncommon to feel angry, helpless and confused, all at the same time.

Naturally, many parents feel a great sense of sadness and grief at the possibility of the death of their child. While it is sometimes difficult to avoid focusing on the possibility of death, it is important to remember that survival rates for children with leukaemia have risen dramatically, and will continue to improve in the future. It is important to remember that the doctors, nurses and other health professionals caring for your child are experts in this area. They have a great deal of knowledge and experience in caring for children with leukaemia.

Every effort will be made to ensure that your child feels comfortable during any test or procedure. For example, local anaesthetic creams may be applied to the skin prior to any necessary needle pricks while stronger painkillers, sedation and/or a general anaesthetic can be given for very painful procedures. If your child requires a general anaesthetic you will be allowed to stay by their side until they are asleep, and be there to greet them again when they wake up afterwards.

Parents are encouraged to stay, where possible, and comfort their child during various tests and procedures. Remaining calm and confident and encouraging your child can be of great assistance during these times. If you find it too distressing you can always stay close by instead, and return to comfort your child as soon as possible afterwards.

It is best for parents to speak directly to their doctor regarding any questions they might have about their child's 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 children with blood cancers.

The Social Work team attached to your treatment centre are available to help you and offer advice on financial assistance that may be available to you in terms of travel, accommodation, and other benefits you or your child may be entitled to. They are also able to offer you emotional support and help with planning your child's care.

### Children

It is not easy to tell a child about a diagnosis of leukaemia. The amount of information that can be given often varies with the child's age and level of intellectual and emotional development. No one knows your child better than you, and no one can tell you when (or how) to tell them about their illness. While very young children are more likely to be concerned about possible separation from a parent, they will need considerable reassurance and comfort, especially in unfamiliar surroundings.

Slightly older children (6-10 years) will have some understanding of the diagnosis. Fear of pain and bodily harm is common in this age group as is the belief that they are in some way responsible for their illness.

Older children and teenagers are generally capable of understanding the implications of their illness. They are usually very concerned about how they look and any potential changes to their appearance can be very worrying. They may also be very concerned about the impact of treatment on their sexual development and fertility. Every opportunity should be given to allow them to express their concerns and to provide them with accurate and relevant information on issues of concern to them.

It is important to allow children of all ages to express their fears and anxieties, to communicate as openly as possible with them and, where appropriate, to include them in decisions regarding their care. In general, it is important to have an open and honest approach, providing children with as much information as they are comfortable with, and that they can understand at the time. In many cases, attempts to withhold information can cause even more anxiety than if the truth had been told from the start.

Many parents find that their child's behaviour regresses while they are sick or in hospital. This is normal. While uncharacteristic behaviours may have gone unchecked during this stressful time, it is important to re-establish rules and boundaries as soon as possible for the child with leukaemia as well as the other children in the family. This will not only contribute to a calmer home environment, it will also help to make the children feel more secure and relaxed.

### Socialising with other children

Interacting with other children is an essential part of any child's social and psychological development. Because of the nature of treatment for leukaemia, most children spend more time out of hospital than in hospital. Between treatments and when your child is well enough they can participate in their usual daily activities including attending playgroups, day care or school. These settings provide children with opportunities for learning, for socialising with their peer group and for making friends. For the child with leukaemia, they can also provide a sense of returning to normal and hope for the future.

## School

Children undergoing treatment from leukaemia may have interrupted school attendance during treatment and at other times when they are unwell. While your child is undergoing treatment it is natural, as a parent, to feel that they may be missing out at school. Be assured that children do catch up. In the meantime they often gain valuable experiences from their time away from school, which can be a special bonding time with parents. Many treatment centres have hospital-based teachers who can help your child stay as up-to-date as possible during these times. In addition, your child's schoolteacher may be able to supply lessons from school, which your child can follow when they feel well enough.

Some children miss their school friends and the social life that comes with being a student. This may be true also for young adults attending university or other training institutions, and for well siblings, where the family has had to relocate for specialist treatment. At times the child or teenager may feel bored, left behind or forgotten about by their friends. Where possible, keeping in contact with the school, informing them of your child's progress and encouraging classmates to keep in contact with your child through visits, phone calls, letters, cards, webcam, Facebook or emails which can be accessed through the hospital. This will benefit them while they are out of school and will also make the transition back to school after or in between treatments easier.

It is important to provide teachers and/or carers an adequate amount of medical information about your child's illness and how the disease or its treatment may affect them at different times. This will help them to anticipate and meet your child's needs. Tiredness and risk of infection are important concerns when your child is undergoing treatment and for some time afterwards. The doctors and nurses at the treatment centre will provide you with information and some common sense strategies to help reduce these risks while allowing your child to lead as normal a life as possible during this time. You can pass this information on to teachers and carers. It is also important to make teachers, carers and other parents aware of your child's situation and the need to be informed about any outbreaks of contagious infections like chicken pox or measles, so that you can take appropriate steps to prevent infection.

Preparing teachers and students for the way your child may look (for example, without their hair), how your child might feel about returning to school (anxious, excited, self-conscious) and how they might make things easier for their classmates (acceptance, inviting them to 'join in') can be important in supporting your child's self confidence and self esteem. When your child does return to school, encourage the teachers and students to treat them as a 'normal' student - just one of the class, while being aware of any special needs they might have.

Many paediatric treatment centres run outreach programs where health professionals, like the oncology liaison nurse, may be able to visit the school and explain the illness both to teachers and to your child's classmates. Educational psychologists, counsellors or school liaison officers can help. Organisations such as Leukaemia & Blood Cancer New Zealand, Child Cancer Foundation, and CanTeen can be a useful source of information and peer support during this time.

Occasionally children experience some learning difficulties as a result of their treatment. Most schools have early intervention and support programs that can assist your child if necessary.

## **Family**

The diagnosis and treatment of leukaemia can cause an extreme amount of stress within any family. The demands of treatment bring many disruptions to normal day-to-day lives. Family routines are often disrupted with frequent trips to the hospital for tests or treatment. Members of the family may suddenly have to perform roles with which they are not familiar, for example cooking, cleaning, and taking care of children. In other cases they may have to take on extra roles and responsibilities within the family, sometimes on top of their paid work. This can be both physically and mentally exhausting.

Some parents find that, where possible, allowing themselves to maintain as much of their familiar role as possible within the family helps to maintain some normality in the situation and give them and everyone else in the family a better sense of control and hope for the future.

Many parents are understandably concerned about the social and financial impact of the diagnosis and treatment of ALL on their families. In many cases one or both parents may have to spend time out of the workforce and away from home while they care for a sick child. There are a variety of programmes designed to help ease the emotional and financial strain created by cancer. Financial support is available through allowances to help with the costs of travel, accommodation and other financial pressures. Practical support is also available from Leukaemia & Blood Cancer New Zealand - contact your local Support Services Coordinator using the details on the back of this booklet. The social worker at your treating hospital will also be able to help you and your family access services from other organisations.

## **Caring for the 'well' sibling**

When a child is diagnosed with cancer the 'well' siblings (brothers and sisters) may experience many confusing emotions. The way in which they respond to these emotions will depend on their age and development level. They may worry about their unwell sibling, and feel sad about family separations. Reassuring siblings that they are loved and giving them opportunities to talk about how they are feeling is important. This helps them to feel better about themselves and acknowledge that what they are feeling is normal and a result of the situation.

During this time all children within the family need a great deal of support, guidance and love. Sticking as much as possible to normal routines like bedtimes, applying the expected boundaries on behaviours and having a reasonable and consistent approach to discipline can help to make children feel more secure, when so many other things appear to be changing within their family.

Giving the sibling appropriate information (and repeating this information when required) about what is happening to the unwell child and including them in some hospital visits can be helpful. This may help to reduce their anxiety and assist them to understand the reasons for the hospital visits and treatment. Asking other family members or friends to spend time with the sibling or take them on a special outing can also help.

CanTeen offers support and activities to patients' siblings who are aged 12 years and over; and the Child Cancer Foundation runs a Siblings' Beads Programme specifically designed to acknowledge the impact that having a brother or sister with cancer has. Contact these organisations via their websites [www.childcancer.org.nz](http://www.childcancer.org.nz) or [www.canteen.org.nz](http://www.canteen.org.nz)

### You and your partner

Serious illness within a family can be very challenging for partner relationships. As well as dealing with the threat of losing a child, treatments make many demands on partners' time and emotional resources.

Effective communication between partners is essential. Acknowledging and talking about the stress in the situation can help. Many treatment centres have a counsellor, psychologist, outreach nurse consultant, social worker and pastoral care workers who can assist you and your family in coping better with the practical and emotional difficulties you may be experiencing. They can also identify strategies that will help you and your family cope during and after treatment.

The Support Services team at Leukaemia & Blood Cancer New Zealand are available to provide you with support and understanding. If necessary they can help to organise counselling for you and your partner.

### Finishing treatment - looking to the future

Once treatment has finished most parents 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 childhood leukaemia. It is important to remember that your treating specialist will send information to your GP to keep them informed regarding your child's progress and what needs to be followed up, on a regular basis, for example blood tests.

Even though your child have been treated successfully for leukaemia it is normal for parents to continue to experience feelings of vulnerability for their child, uncertainty about the future and fear that the illness could return.

The fear of a recurrence or relapse of leukaemia may cause some parents to become overprotective of their child. Naturally, they are more aware of any physical signs and symptoms than previously. For example, a bruise, which the child has sustained in normal play, may cause the parent to become very anxious that this may be a sign that their child has relapsed.

Follow-up appointments after treatment has finished are often times of great anxiety as people wait for the '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 their child's illness.

Many people find it useful to talk with other parents and family members who understand the complexity of feelings and the kinds of issues that come up for parents and families living with an illness of this nature. Support groups can offer important information and a supportive environment for people to discuss issues important to them. Leukaemia & Blood Cancer New Zealand have information about relevant support groups available in your area.

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](http://www.lifebloodlive.org.nz).

## 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](http://www.leukaemia.org.nz)  
[www.lifebloodlive.org.nz](http://www.lifebloodlive.org.nz)

### **Child Cancer Foundation**

[www.childcancer.org.nz](http://www.childcancer.org.nz)

### **CanTeen**

[www.canteen.org.nz](http://www.canteen.org.nz)

### **Make a Wish Foundation**

[www.makeawish.org.nz](http://www.makeawish.org.nz)

### **Grief Centre**

[www.griefcentre.org.nz](http://www.griefcentre.org.nz)

### **Skylight Foundation**

[www.skylight.org.nz](http://www.skylight.org.nz)

### **Cancer Society of New Zealand**

[www.cancernz.org.nz](http://www.cancernz.org.nz)

### **Leukaemia Foundation of Australia**

[www.leukaemia.org.au](http://www.leukaemia.org.au)

### **MacMillan Cancer Support (A UK cancer information site)**

[www.macmillan.org.uk](http://www.macmillan.org.uk)

### **Leukemia & Lymphoma Society of America**

[www.leukemia-lymphoma.org](http://www.leukemia-lymphoma.org)

### **Leukaemia & Lymphoma Research Fund (UK)**

[www.llresearch.org.uk](http://www.llresearch.org.uk)

## 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 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 other substances such as bacteria, viruses and some cancer cells and cause their destruction.

### **Antibiotic**

A drug used to prevent or treat bacterial infections.

### **Antiemetic**

A drug used to prevent or reduce feelings of sickness (nausea) and vomiting.

### **Anti-fungal**

A drug used to prevent or treat fungal infections.

### **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. 'Antigen' is also the general term used to describe proteins found on the surface of all body cells. Here, antigens act like flags identifying different types of cells.

### **B-cell**

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

### **Blast cells**

Immature blood cells normally found in the bone marrow. Blast cells normally constitute up to 5 per cent of all bone marrow cells. These cells divide and replenish all the normal blood cells in the marrow and circulating blood. Acute leukaemia is characterised by an accumulation of abnormal blast cells that take over the marrow and spill out into the blood stream.

### **Blood count**

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

### **B-lymphocyte (B-cell)**

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

**Bone marrow biopsy**

A procedure to collect a sample of the bone marrow. This is usually from the back of the hip bone, or occasionally from the breastbone (sternum). This procedure usually done under general anaesthetic for children and incorporates either or both of the following:

Aspirate – A procedure that involves removing (or aspirating) a small sample of bone marrow fluid for examination in the laboratory.

Trephine – A procedure that involves removing a small core of bone and bone marrow for examination in the laboratory.

**Cancer**

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

**Cannula**

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

**Central venous catheter (CVC)**

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

**Cerebrospinal fluid (CSF)**

The fluid that surrounds and protects the brain and spinal cord. Samples of this fluid can be collected for examination using a procedure known as a 'lumbar puncture'. Chemotherapy is sometimes given into the cerebrospinal fluid to prevent or treat cancer in the central nervous system (CNS).

**Chemotherapy**

Single drugs or combinations of drugs which may be used to kill and prevent the growth and division of cancer cells. Although aimed at cancer cells, chemotherapy can also affect rapidly dividing normal cells and this is responsible for some common side effects including hair loss and a sore mouth. Nausea and vomiting are also common, but nowadays largely preventable with modern anti-nausea medication. Most side effects of are temporary and reversible.

**Chromosomes**

Chromosomes are made up of coils of DNA (deoxyribonucleic acid). DNA carries all the genetic information for the body in sequences known as genes. There are approximately 40,000 genes on 23 different chromosomes. The chromosomes are contained within the nucleus of a cell.

**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 5 per cent. There are no blast cells present in the circulating blood and the blood count has returned to normal.

**Computerised axial tomography (CT scan or CAT scan)**

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

**Cortico-steroids (steroids)**

A group of man-made hormones including prednisone, prednisolone, methylprednisolone and dexamethasone used in the treatment of certain blood and bone marrow cancers. As well as having anti-cancer effects, cortico-steroids also have anti-inflammatory and immunosuppressive (anti-rejection) effects.

**Cure**

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

**Cytogenetic tests**

The study of the genetic make-up of the cells, in other words, the structure and number of chromosomes present. Cytogenetic tests are commonly 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**

Where the disease is getting worse on or off treatment.

**DNA (Deoxyribonucleic acid)**

Molecules found in the center of the cell that carry all the genetic information for the body. There are four different chemical compounds of DNA (bases) arranged in coded sequences called genes, which determine an individual's inherited characteristics.

**Genes**

Collections of DNA. Genes direct the activity of cells. They are responsible for the inherited characteristics that distinguish one individual from another.

**Growth factors**

A complex family of proteins produced by the body to control the growth, division and maturation of blood cells by the bone marrow. Some are now available as drugs as a result of genetic engineering and may be used to stimulate normal blood cell production following chemotherapy or bone marrow or peripheral blood cell transplantation. For example G-CSF (granulocyte colony stimulating factor).

**Haemopoiesis**

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.

**High-dose therapy**

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

**Immune system**

The body's defence system against infection and disease.

**Immunophenotyping**

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

**Intrathecal injection**

Injection of drug(s) into the cerebrospinal fluid (CSF) (the fluid that surrounds the brain and spinal cord). The space between the brain and spinal cord and their coverings is known as the intrathecal space.

**Late effects**

Side effects of chemotherapy and/or radiotherapy that may only become apparent with long-term monitoring over a period of years.

**Leukaemia**

A 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 take over the bone marrow often causing a fall in blood counts. If they spill out into the bloodstream however they can cause very high abnormal white cell counts.

**Leukaemic blasts**

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

**Lumbar puncture**

A procedure used to remove fluid from around the brain and spinal cord (cerebrospinal fluid or CSF) for examination in the laboratory. A lumbar puncture may also be used to administer chemotherapy into this fluid to prevent or treat disease in the central nervous system (CNS).

**Lymph nodes or glands**

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

**Lymphatic system**

A vast network of vessels, similar to blood vessels, that branch out into all the tissues of the body. These vessels carry lymph, a colourless watery fluid that carries lymphocytes, specialised white cells that protect us against disease and infection. The lymphatic system is part of the body's immune system.

**Lymphocytes**

Specialised white cells that help defend 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.

**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.

**Malignancy**

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

**Menopause**

The stopping of menstruation (periods). Also called 'the change of life'.

**Mucositis**

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

**Mutation**

A change in the DNA code of a cell, caused for example by exposure to hazardous chemicals or copying errors during cell division. If mutations affect normal cell function this can lead to the development of disease due to the loss of normal function or the development of abnormal functions of that cell.

**Myeloablative therapy**

High dose chemotherapy or radiotherapy used to destroy disease but which also destroys the patient's own bone marrow. A stem cell transplant is needed to restore normal bone marrow function following myeloablative therapy.

**Myeloid**

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

**Neutropaenia**

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

**Neutrophils**

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

**Philadelphia chromosome**

The abnormal chromosome present in nearly all cases of chronic myeloid leukaemia and some cases of acute lymphoblastic leukaemia. It 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.

**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**

The disease is not responding to treatment.

**Remission**

When there is no evidence of disease detectable in the body. This is not the same as a cure as relapse may still occur.

**Spleen**

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

**Standard therapy**

The most effective and safest therapy currently being used.

**Stem cells**

Stem cells are primitive blood cells that can give rise to more than one cell type. There are many different types of stem cells 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**

General name given to bone marrow and peripheral blood stem cell transplants. These treatments 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 serious diseases.

**T-Cell**

A type of white cell involved in controlling immune reactions.

**Translocation**

A chromosomal abnormality in which part of the one chromosome is transferred to another.

**White cells**

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

**X-ray**

A form of radiation used in diagnosis and treatment.

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

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

- Dictionary of Terms
- Acute Lymphoblastic Leukaemia in Adults
- Haematology Patient Diary
- Acute Lymphoblastic Leukaemia in Children
- Clinical Trials
- Acute Myeloid Leukaemia
- Autologous Stem Cell Transplants
- Chronic Lymphocytic Leukaemia
- Allogeneic Stem Cell Transplants
- Chronic Myeloid Leukaemia
- Myeloproliferative Disorders
- Hodgkin Lymphoma
- Myelodysplastic Syndromes
- Non-Hodgkin Lymphoma
- Myeloma
- 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
- Leukaemia Today
- Lymphoma 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](mailto: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](mailto:info@leukamia.org.nz)

**Notes**

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**Acute Lymphoblastic Leukaemia in Children**

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?**

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

Yes     No

If yes, what were they?

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


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**6. What did you like least about this booklet?**


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**7. Any other comments?**


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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



our mission is to care, our vision is to cure

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**Telephone** 09 638 3556  
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**[leukaemia.org.nz](http://leukaemia.org.nz)**

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