

Understanding Acute Lymphoblastic Leukaemia (ALL) in Children

A guide for parents and families



Leukaemia
Foundation[®]

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CONTENTS

	PAGE
Acknowledgements	2
Introduction	3
The Leukaemia Foundation	4
Bone marrow, stem cells and blood cell formation	8
The lymphatic system	14
What is leukaemia?	15
What is acute lymphoblastic leukaemia (ALL)?	17
How common is ALL and who gets it?	17
What causes ALL?	18
What are the symptoms of ALL?	19
How is ALL diagnosed?	20
Which type of ALL does my child have?	23
How is ALL treated?	25
Types of treatment	28
Phases of treatment	31
Side effects of treatment	36
Shared care	42
Follow up	42
Long-term effects of treatment	43
Complementary therapies	45
Nutrition	45
Making treatment decisions	46
Social and emotional issues	47
Useful internet addresses	55
Glossary of terms	56

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The 2010 revisions were provided by Dr Margaret Little and Bernadette O'Grady and approved by Professor Ken Bradstock. The Leukaemia Foundation values feedback from patients, their families, carers and health care professionals working with people affected by blood disorders. If you would like to make suggestions, or tell us about your experience of using this booklet, please contact the National Manager, Support Services at info@leukaemia.org.au.

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INTRODUCTION

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

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. Remember that this is a general booklet and not everything written here will necessarily apply to you and your child's experience of leukaemia. It is not the intention of this booklet to recommend any particular form of treatment to you. You need to discuss your circumstances at all times with your doctor and treatment team.

We have used some medical words and terms, which you may not be familiar with. These are highlighted in *italics*. Their meaning is explained in the booklet or in the glossary of terms at the back of the booklet.

In some parts of the booklet we have provided additional information you may wish to read on selected topics. This information is presented in the shaded boxes. Some of you 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 treating hospital.

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.

Finally, we hope that you find this information useful and we would appreciate any feedback so that we can continue to serve you and your families better in the future.

THE LEUKAEMIA FOUNDATION

The Leukaemia Foundation is the only national not-for-profit organisation dedicated to the care and cure of patients and families living leukaemias, lymphomas, myeloma and related blood disorders. Since 1975, the Foundation has been committed to improving survival for patients and providing much needed support. The Foundation does not receive direct ongoing government funding, relying instead on the continued and generous support of individuals and corporations to develop and expand its services.

The Foundation provides a range of **free** support services to patients and their carers, family and friends. This support may be offered over the telephone, face to face at home, hospital or at the Foundation's accommodation centres, depending on the geographical and individual needs. Support may include providing information, patient education seminars and programs that provide a forum for peer support and consumer representation, practical assistance, accommodation, transport and emotional support/counselling.

The Leukaemia Foundation funds leading research into better treatments and cures for leukaemias, lymphomas, myeloma and related blood disorders. Through its National Research Program, the Foundation has established the PwC Leukaemia and Lymphoma Tissue Bank based at the Princess Alexandra Hospital in Brisbane, and the Leukaemia Foundation Research Unit at the Queensland Institute for Medical Research. In addition, the Foundation also funds research grants, scholarships and fellowships for talented researchers and rural health professionals.

Support Services



"Foundation staff provide patients and their families with information and support across Australia"

The Leukaemia Foundation has a team of highly trained and caring Support Services staff with qualifications and/or experience in nursing and allied health that work across the country. They can offer individual support and care to you and your family when it is needed.

The support services may include:

Information

The Leukaemia Foundation has a range of booklets (such as this one), fact sheets and other resources that are available free of charge. These can be ordered via the form at the back of this booklet or downloaded from the website. Translated versions (in languages other than English) of some booklets and fact sheets are also available from our website.

Education & support programs

The Leukaemia Foundation offers you and your family disease-specific and general education and support programs throughout Australia. These programs are designed to empower you with information about various aspects of diagnosis and treatment and how to support your general health and well being.

Emotional support

A diagnosis of a blood cancer/disorder can have a dramatic impact on a person's life. At times it can be difficult to cope with the emotional stress involved. The Leukaemia Foundation's Support Services staff can provide you and your family with much needed support during this time. They may refer you or a loved one to a specialist if professional counselling services are required.

Online discussion forum

The Foundation has established an on-line information and support group for people living with leukaemia, lymphoma, multiple myeloma, or a related blood disorder. Registration is free and participants can remain anonymous, see www.talkbloodcancer.com

Accommodation

Some patients and carers need to relocate for treatment and may need help with accommodation. The Leukaemia Foundation staff can help you to find suitable accommodation close to your hospital or treatment centre. In many areas, the Foundation's fully furnished self-contained units and houses can provide a 'home away from home' for you and your family.

Transport

The Foundation also assists with transporting patients and carers to and from hospital for treatment. Courtesy cars and other services are available in many areas throughout the country.

Practical Assistance

The urgency and lengthy duration of medical treatment can affect you and your family's normal way of life and there may be practical things the Foundation can do to help. In special circumstances, the Leukaemia Foundation provides financial support for patients who are experiencing financial difficulties or hardships as a result of their illness or its treatment. This assistance is assessed on an individual basis.

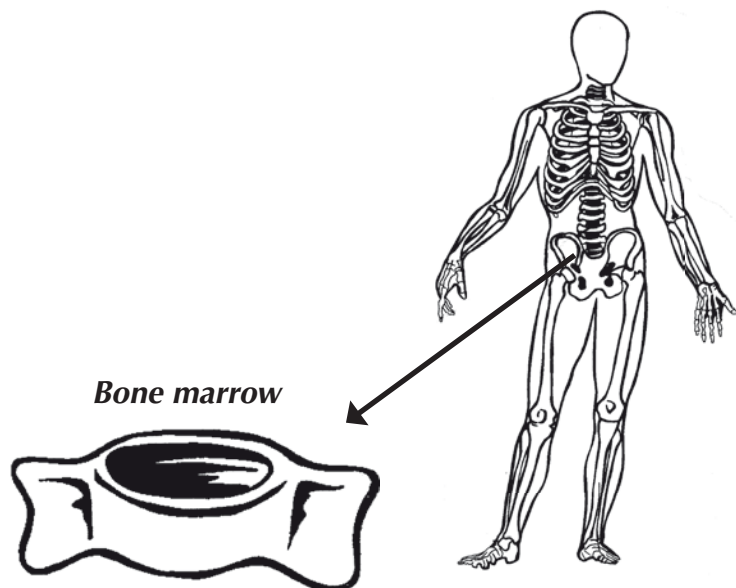
Young Adults

A website for young adults has been developed called “Revive”. This site has information specifically designed for young adults and contains a discussion forum to allow patient to patient interaction and support. The site is www.teamrevive.com

Contacting us

The Leukaemia Foundation provides services and support in every Australian state and territory. Every person’s experience of living with these blood cancers and disorders is different. Living with leukaemias, lymphomas or myeloma is not easy, but you don’t have to do it alone. Please call **1800 620 420** (Freecall) to speak to a local support service staff member or to find out more about the services offered by the Foundation. Alternatively, contact us via email by sending a message to info@leukaemia.org.au or visit www.leukaemia.org.au

BONE MARROW, STEM CELLS AND BLOOD CELL FORMATION



Bone marrow

Bone marrow is the spongy tissue that fills the cavities inside your bones. Most of your blood cells are made in your bone marrow. The process by which blood cells are made is called *haemopoiesis*.

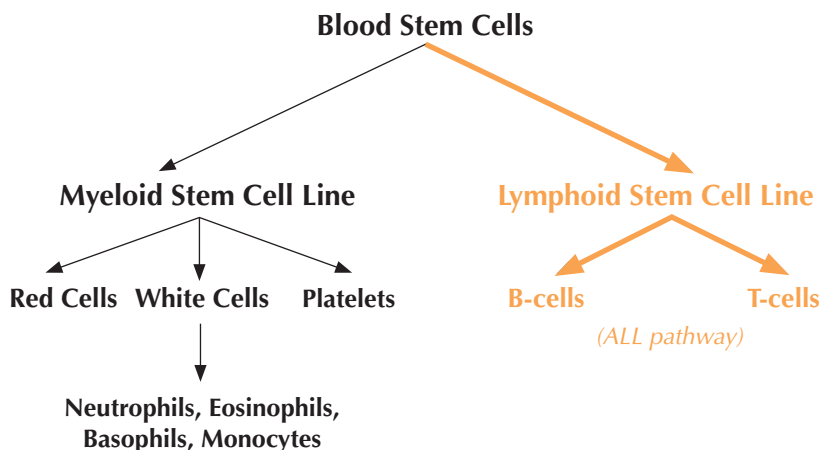
As an infant, haemopoiesis takes place at the centre of all bones. In later life, it is limited to the hips, ribs and breastbone (*sternum*). Some of you may have had a *bone marrow biopsy* taken from the bone at the back of your hip (the *iliac crest*) or the breastbone.

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 small in number but are able, when stimulated, to reproduce vast numbers of red cells, white cells and platelets. All blood cells need to be replaced because they have limited life spans.

There are two main families of stem cells, which develop into various types of 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-cells and B-cells.



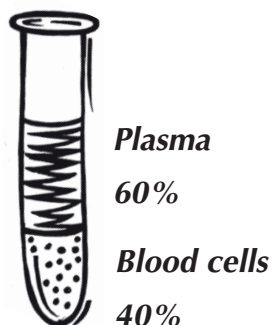
Growth factors and cytokines

All normal blood cells have a limited survival in 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.

These days some 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.

Blood



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

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 will be given a copy of your child's blood results, which should 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.

Normal range of blood values for children

	1 month	1 year	3 years	5 years	9 years	16 years
Haemo-globin g/L	102-130	104-132	107-136	110-139	113-143	115-165 F 130-180 M

Anaemia

Anaemia is a condition caused by a reduction in the number of red cells or 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 because they are not getting enough oxygen. In this situation a red cell transfusion may be given to restore the red cell numbers and therefore the haemoglobin to more normal levels.

White cells

White cells fight infection. There are different types of white cells that fight infection together and in different ways.

Roles of white cells

<i>Neutrophils</i>	<i>kill bacteria and fungi</i>
<i>T-Cells</i>	<i>kill viruses, parasites and cancer cells; produce cytokines</i>
<i>B-Cells</i>	<i>make antibodies which target microorganisms</i>
<i>Monocytes</i>	<i>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</i>
<i>Eosinophils</i>	<i>kill parasites.</i>
<i>Basophils</i>	<i>work with neutrophils to fight infection.</i>

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
White cell count x $10^9/L$	6.4-12.1	5.4-13.6	4.9-12.8	4.7-12.3	4.7-12.2	3.5-11

Neutropenia

Neutropenia is the term given to describe a lower than normal neutrophil count. If your child is neutropaenic (neutrophil count of less than $1 \times 10^9/L$) they are considered to be at risk of developing frequent and sometimes severe infections.

Normal neutrophil count for children:

	1 month	1 year	3 years	5 years	9 years	16 years
Neutrophils x $10^9/L$	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
Platelets x $10^9/L$	270-645	205-553	214-483	205-457	187-415	150-450

Thrombocytopenia

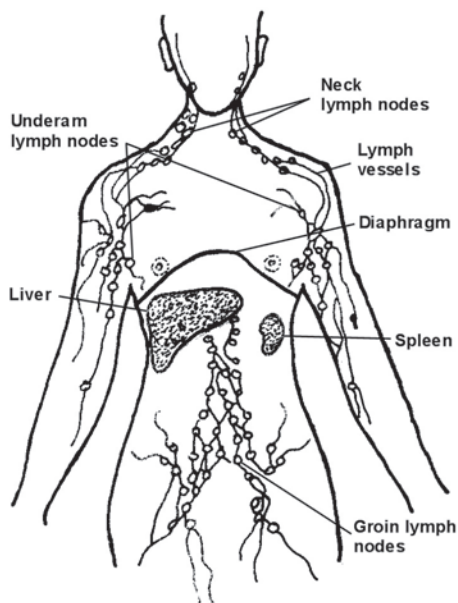
Thrombocytopenia is the term used to describe a reduction in the platelet count to below normal. If your child's platelet count drops below 10 ($10 \times 10^9/\text{L}$) they are at risk of bleeding and tend to bruise easily. Platelet transfusions are sometimes given to bring the platelet count back to a safe level.



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-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 that live there 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.

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

Acute leukaemias

Acute leukaemias develop and progress quickly and therefore need to be treated as soon as they are diagnosed. Acute leukaemias affect very immature blood cells, preventing them from maturing properly.

Chronic Leukaemias

In **chronic leukaemias** there is an accumulation of more mature but abnormal white cells. Chronic leukaemias can occur at all ages but they are rarely seen in children.

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 (see diagram on page 9).

Myeloid leukaemias

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

Lymphoid leukaemias

When leukaemia starts somewhere in the lymphoid stem cell line it is called lymphoblastic, lymphocytic, or lymphatic leukaemia (see diagram on page 9).

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. CML is very rare in children and CLL virtually never occurs in this age group. ALL is the most common leukaemia in children and AML occurs occasionally.

* There are separate Leukaemia Foundation booklets that provide more details about these diseases.

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

Improvements in the diagnosis and treatment of children with ALL mean that, these days, almost all children treated for ALL will achieve a remission from their disease and most will be cured.

HOW COMMON IS ALL AND WHO GETS IT?

Each year in Australia around 225 children are diagnosed with leukaemia. Of these around 175 children are diagnosed with acute lymphoblastic leukaemia (ALL), making it the most common type of cancer overall in children aged 0 to 14 years. The incidence of ALL is highest in children between the ages of 2 and 4 years. It is more commonly diagnosed in boys.

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

* There are separate Leukaemia Foundation booklets that provide more details about these diseases.

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 going on all the time into possible causes and a number of environmental factors continue to be investigated. To date however, 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.

Infections

There is some evidence to suggest that viral infections may play a role in the development of ALL in some children. 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 however contagious. A child cannot 'catch' ALL by being in contact with someone who has it.

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 however unlikely that any children born in Australia 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 Australia however are unlikely to be exposed to high enough levels of these chemicals to cause ALL.

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 however 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 and Fanconi's anaemia are at an increased risk of developing ALL.

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 your child to have 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-kee-a') 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

Bone and / or joint pain is common and results from the marrow being literally "stuffed" with leukaemic cells. Occasionally there may be deposits of leukaemic 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. So, 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 child's blood and bone marrow.

Full blood count

The first step in diagnosing ALL requires a simple blood test called a *full blood count* (FBC) or *complete blood count* (CBC). This involves taking a sample of your 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 (anaemia), and a low platelet count. Most children have a high white cell count and almost all children will have abnormal leukaemic blast (immature) 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 a repeat bone marrow examination to assess how well their 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* and *cytogenetic* tests.

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

This test uses special markers called antigens found on the surface of blast cells to determine the exact subtype of leukaemia your child has and therefore the best way to treat it.

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 your child's disease originated (B-cell ALL or T-cell ALL), and the point at which this cell stopped developing properly in the bone marrow.

Cytogenetic ('cy-to-gen-etic') **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.

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.

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

Cerebrospinal fluid examination

A small sample of the cerebro-spinal fluid (CSF) that surrounds your child's brain and spinal cord is collected, during a procedure called a lumbar puncture which is usually performed under a short general anaesthetic or local anaesthetic. This is then tested in the laboratory to check for the presence of cancer cells within the central nervous system (CNS). If cells are found, additional treatment is given to the CNS. The CSF is usually negative but even in this case some protective CNS directed treatment (CNS prophylaxis) is required as a very small number of cells may be present but not detectable.

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, x-rays and ultrasound. Blood tests may include kidney function tests, liver function tests and coagulation tests, to see if your child's blood is clotting properly.

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 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 stem cell line in the bone marrow. Depending on the main type of abnormal lymphocyte present, ALL can be broadly classified into two main groups:

- ALL that arises in developing B-cells and
- ALL that arises in developing T-cells.

Some years ago doctors from America, France and Britain decided to classify ALL into three different subtypes (L1, L2 and L3) based on the appearance of the leukaemic cells under the microscope (*morphology*). Each subtype provides information on the type of blood cell involved and the point at which it stopped maturing properly in the bone marrow. This is known as the French-American-British (FAB) classification system.

The current World Health Organisation's classification system for ALL uses additional information, obtained from more specialised laboratory techniques, like immunophenotyping and cytogenetic tests (see above), to classify ALL more precisely. The diagnosis of different subtypes of ALL depends on the detection of distinct cell surface markers (CD antigens), some of which correspond with normal lymphocytes in various stages of development. The leukaemic cells are however recognisably different from normal lymphocytes due to differences in their size, structure and how they look under the microscope.

Pre-B-cell ALL

In around 80% of cases, childhood ALL arises in 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 non-Hodgkin 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 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).

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 referral 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) 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 Australia who are diagnosed with ALL usually follow established protocols or plans of treatment as part of large national or 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.

It is important to realise that whatever protocol your child follows, it will be the best treatment known against ALL, 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.

Informed consent

Giving an 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.

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.

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 they are following.

Chemotherapy is usually given as a combination of drugs (combination chemotherapy). These drugs act together and in different ways to destroy the leukaemic cells. Chemotherapy is usually given in several cycles (or courses) with rest periods in between. This is to allow your child's body (the bone marrow in particular) time to recover from the side-effects.

Chemotherapy is given in many different ways in the treatment of ALL. Some drugs are given in tablet 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 (IT or into the spinal fluid), 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.

Most of the time your child will not need to be admitted to hospital for chemotherapy. After their initial treatment they may be able to receive a majority of the rest of their treatment in the outpatient's department of the hospital or clinic, or at home. Sometimes however, depending on the type of chemotherapy being given or your child's general health, they may need to be admitted to hospital.

Corticosteroid therapy

Corticosteroids are hormones produced naturally by the body. They can also be made in the laboratory. These drugs play an important role in the management of leukaemia. *Prednisone*, *prednisolone* and *dexamethasone* are examples of corticosteroids 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 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 (intrathecal injection), through a lumbar puncture. Some types of intravenous chemotherapy and corticosteroid 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 the disease is found here at diagnosis no additional treatment is required. Your child's haematologist / oncologist 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.

Commonly Used Terms

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. For many others treatment can help to control their disease for a long time.

Complete remission

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.

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 re-appear (relapse) over time.

Resistant or refractory disease

This means that the leukaemia is not responding to treatment.

Relapse

The leukaemia has re-appeared. 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.

PHASES OF TREATMENT

Treatment for ALL can be divided into three phases:

- Remission induction therapy
- Consolidation therapy (including interim maintenance and intensification)
- Maintenance therapy

Depending on the protocol your child is following, remission induction, consolidation, interim maintenance and intensification phases of treatment can last for up to 10 months.

Remission induction therapy

Soon after your child is diagnosed they will need to begin an intensive course of treatment to bring about, or *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 this first phase of treatment.

Commonly used chemotherapy drugs in this phase of treatment include: vincristine, daunorubicin, asparaginase and dexamethasone (corticosteroid). CNS therapy also begins at this stage.

While your child is having 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. In patients where there is a high risk of this complication (such as very high leukaemia cell count) a new drug called *rasburicase* may be used to protect the kidneys. High volumes of fluid are also given intravenously to help flush through the kidneys.

Almost all children with ALL will achieve a remission following induction therapy. In a small number of cases however, the disease does not respond to treatment as expected and 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 therapy (intensification)

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 re-appearing (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 (see below). Consolidation therapy usually involves 'blocks' of intensification treatment over several months and includes additional drugs such as cyclophosphamide, cytarabine, etoposide and thioguanine. This is given to reduce left over disease to a minimum (minimum residual disease).

Risk-based therapy

A prognosis is an estimate of the likely course of a disease and whether it is likely to relapse in the future. It provides some guide regarding the chances of curing the disease or controlling it for a given time. 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. In many protocols a one-week course of corticosteroids is given (on its own) to reduce the amount of leukaemia with minimal side effects. The response to this "steroid prophase" is an important factor in determining your child's prognosis.

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 has spread to the 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 significant proportion (20-25%) will experience a relapse over time.

Minimal residual disease

We now know that there is usually a strong relationship between the number of leukaemic cells left over 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 left over 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.

Maintenance therapy

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 corticosteroids 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 2 years for girls and just over 3 years for boys. During this time your child will be treated as an outpatient.

As soon as they are well enough, children are encouraged to take part in their usual daily activities including attending school or day care. 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.

Haemopoietic stem cell transplantation

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 but now the source of cells may be from marrow or 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 haemopoietic 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 is a separate Leukaemia Foundation booklet called 'Understanding Allogeneic Transplants - A guide for patients and families' that provides more details on this type of treatment.*

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 haemopoietic 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. For relapsed or refractory ALL in children, it is fortunate that there are some new therapies available.

SIDE-EFFECTS OF TREATMENT

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.

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 your child's blood) will generally fall within a week of treatment, increasing their risk of infection and bleeding.

Neutropenia

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), avoiding other children who have recently had a live vaccine like 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. In general there is no need for your child to stop going to school or playgroup at any stage during treatment provided they feel up to it. As a precaution, family members may be advised to have the flu vaccination. They will not contract serious infections at school (provided sensible precautions are taken by avoiding classmates with chicken pox) and the benefits of maintaining social contacts outweigh any disadvantages. Some centres advise not to attend school/daycare until after induction therapy. 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.

Thrombocytopenia

Your child's platelet count may also be affected by their disease and by the chemotherapy they are receiving and 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 chop bones as these can 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. Taking the child's temperature rectally, or the insertion of rectal suppositories should also be avoided for the same reasons. Your child is likely to 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.

Anaemia

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.

When to call the doctor ...

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

- **a temperature of 38.5°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 your 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 venous catheter site.

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

Hair loss

Hair loss is a very common side-effect of chemotherapy and some forms of radiotherapy. It is, however, usually only temporary. The hair starts to fall out within a couple of weeks of treatment and 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 bandanas. Older children and teenagers

however, 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.

As well as seeking supportive counselling from relevant members of the treatment team, teenagers may find it useful to talk with other teenagers who understand the complexity of feelings and the kinds of issues that come up for young people living with an illness of this nature. This can be arranged by contacting CanTeen, the national support organisation for young people (aged 12 – 24 years) living with cancer. CanTeen can be contacted by telephoning 1800 226 833, or by visiting their website at: www.canteen.org.au. The Leukaemia Foundation has an information booklet for young people living with a blood cancer. Contact your local support services coordinator to obtain a copy.

Mucositis

Mucositis, an 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. After this they may be given some medication to help stop the diarrhoea and relieve any discomfort they may be feeling.

Your child's bottom can become quite sore if they have diarrhoea. 'Baby wipes' are a good idea for cleaning their bottom at this time because 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 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 having 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.*

Seizures

Intrathecal (IT) therapy is rarely associated with seizures, otherwise known as fitting. If your child experiences a seizure, or if the doctor feels they may be at risk of having a seizure, they will prescribe special medication to help to prevent this from happening.

Side-effects of corticosteroids

Side-effects of corticosteroids depend largely on how long they are used for, and the dose given. Again, children respond differently. An increased appetite, fluid retention and 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 corticosteroids and frequently want to eat around-the-clock. 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 may require some night sedation. 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, maintaining 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 unpleasant longer-term behavioural problems, which can cause considerable stress within any family.

**There is a separate Leukaemia Foundation booklet called 'Eating Well - A practical guide for people living with leukaemias, lymphomas and myeloma' that provides more information on nutrition during this time.*

Long-term use of steroid therapy may cause other effects such as fluid retention, an increased susceptibility to infections or osteoporosis, where the bones may become weak and brittle. These effects are not common however as most children with ALL do not require prolonged steroid therapy.

Remember to tell your doctors and nurses about any symptoms 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 severe pneumonia. Treatment with Cotrimoxazole generally continues until chemotherapy is completed.

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 at your local children's unit. This may just be regular blood checks, or range from 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 unit to ensure that both are kept up to date with all that is happening with your child.

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 the 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 CNS (now only rarely used in ALL) can cause a number of long-term problems including obesity, reproductive difficulties (discussed below) and delayed growth. Delayed growth can be treated using growth hormone (GH) 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 most commonly seen in younger children. Your child's school progress is monitored as part of their routine follow up after treatment.

Fertility

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 the child when they were treated and the type of treatment they received. In boys, sperm production may be impaired for a while following chemotherapy but it is important to realise that 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. The onset of puberty can also be affected and some children may require hormone supplements to ensure normal sexual development.

Preserving fertility

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. You should discuss sperm banking with your doctor before your son starts any treatment that might impact on his fertility. In some cases however, 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.

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 program) but cannot be undertaken in young children. Unless a haemopoietic 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 remains 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.

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 achieve the best outcomes for children with ALL while reducing the risk and impact of any long-term effects of treatment.

COMPLEMENTARY THERAPIES

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

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 cause any problems with the disease, or its medical treatment.

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.

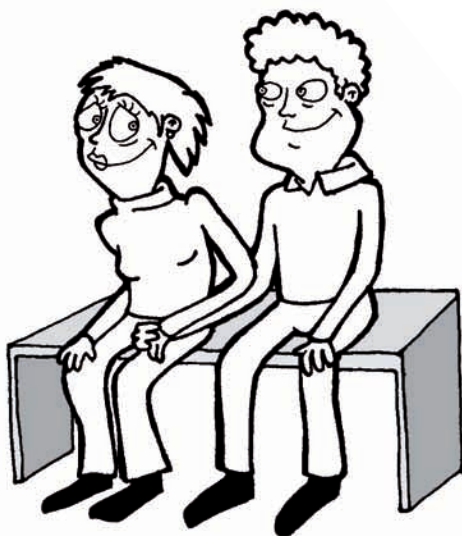
**There is a separate Leukaemia Foundation booklet called 'Eating Well: a practical guide for people living with leukaemias, lymphomas, myeloma and related blood disorders'.*

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 is 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 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 is handy to keep a notebook or some paper and a pen handy as many questions are thought of in the early hours of the morning.

Sometimes it is hard to remember everything the doctor has said. It 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. It is important however 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 organize the weeks ahead.

SOCIAL AND EMOTIONAL ISSUES

Parents

Parents cope with a diagnosis of childhood leukaemia in different ways and there is no right or 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.

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 you and 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 leukaemia treatment 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 children, where the family has had to relocate for specialist treatment. At times the child or adolescent 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 from class mates, cards or posters with thoughtful messages, webcam, videos 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 with 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 put them in a better position 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 steps to prevent your child from infection.

Preparing teachers and students for the way your child may look (for example without their hair) and how they might feel about returning to school (anxious, excited, self-conscious) and how they might make things easier for their classmate (for example 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 like CanTeen, the Make-a-Wish Foundation and the Starlight Foundation can be a useful source of information and peer support during this time. Ask the Leukaemia Foundation for further information about help available to you and your child.

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

The 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, doing the banking 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.

Relocating to hospital for treatment

Treatment for childhood leukaemia, especially in the early stages, requires specialist care that is usually only available at metropolitan hospitals. 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. Many treatment centres now have reasonably priced accommodation for you and your family, on site or close to the hospital. Suitable accommodation can be arranged by contacting the social worker at your treatment centre even before you leave home. The social worker can also tell you about any government assistance schemes (like the Patient Assisted Travel Schemes or PATS) that can provide financial assistance for your travel and accommodation costs. They can also assist you with any paperwork required when making claims for financial assistance. In some areas the Leukaemia Foundation has accommodation for

patients and families. Contact the social worker or the Leukaemia Foundation in your state for more information.

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 programs designed to help ease the emotional and financial strain created by cancer. Financial support is available through pensions and benefits to help with the costs of travel, accommodation and some drugs. Financial counselling is also available free of charge from many charitable organisations, including the Leukaemia Foundation. Financial and practical support is also available from the Leukaemia Foundation and several other organisations including Redkite. The social worker at your treating hospital will be able to help you and your family access these services.

Caring for the ‘well’ sibling

When a child had been diagnosed with cancer the ‘well’ siblings (sisters and brothers) 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 the sick 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 sick 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.

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 staff at the Leukaemia Foundation are there 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 people 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 him or her informed regarding your child's progress and what needs to be followed up, on a regular basis, for example blood tests.

Even though their children 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 their 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 an 'all clear' from their doctor. As time passes and as more distance is allowed between appointments anxiety reduces. Everyone gradually becomes more and more engaged in the activities of daily living rather than concentrating most of their attention on the experience of 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. Ask your doctor or nurse if your treating hospital runs a support group, which might be suitable. If not, they may be able to provide you with the details of a group being run in your area. The Leukaemia Foundation will also have information about relevant support groups.

The Leukaemia Foundation is always here to provide you and your family with information and support to help you cope during this time. Contact details for your state office of the Leukaemia Foundation are provided on the back of this booklet.

USEFUL INTERNET ADDRESSES

- Leukaemia Foundation
www.leukaemia.org.au
- American Cancer Society
www.cancer.org
- Arrow Foundation
www.arrow.org.au
- Australian Bone Marrow Donor Registry
www.abmdr.org.au
- Bone & Marrow Transplant Information Network
www.bmtinfonet.org
- CancerBACUP (A UK cancer information site)
www.cancerbacup.org.uk
- Cancer Council of Australia
www.cancercouncil.com.au
- CanTeen,
www.canteen.org.au
- Centre for Grief and Loss
www.grief.org.au
- Leukaemia Foundation's Network for Young Adults
www.teamrevive.org
- Leukaemia Foundation's Online Forum
www.talkbloodcancer.com
- Leukemia & Lymphoma Society of America
www.leukemia-lymphoma.org
- Leukaemia Research Fund (UK)
www.leukaemia-research.org.uk
- Look Good ... Feel Better program
www.lgfb.org.au
- Make-a-Wish Foundation of Australia®
www.makeawish.org.au
- National Cancer Institute (USA)
www.cancer.gov/cancerinfo
- Redkite (previously the Malcolm Sargent Cancer Fund for Children)
www.redkite.org.au
- Starlight Children's Foundation of Australia
www.starlight.org.au

GLOSSARY OF TERMS

Acute leukaemias

Rapidly progressing cancers of the blood and bone marrow, usually of sudden onset and characterised by uncontrolled growth of immature blood cells which crowd the bone marrow and spill out into the bloodstream.

Acute lymphoblastic leukaemia (ALL)

A rapidly progressing cancer of the blood and bone marrow. ALL affects the type of developing white blood cells known as lymphocytes. It is the most common form of childhood leukaemia, and the most common type of childhood cancer. It also occurs in adults.

Alopecia

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

Allogeneic stem cell transplant

The transplant of blood stem cells from one person to another. The donor is usually a sister or brother or an unrelated volunteer donor.

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-lymphocyte (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). A routine blood test that measures the number and type of cells circulating in the blood.

Blood stem cells

Primitive blood-forming cells that normally live in the bone marrow. They divide and mature into all the different types of blood cells (red cells, white cells and platelets), including the cells of our immune system.

B-cell

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

Bone marrow

The tissue found at the center 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 aspirate

A procedure that involves removing a small sample of bone marrow fluid for examination in the laboratory. The fluid is drawn, under local or general anaesthetic, usually from the back of the hip, or occasionally from the breastbone.

Bone marrow biopsy

A procedure that involves removing a small core of bone marrow for examination in the laboratory. The biopsy (or trephine) is taken under local or general anaesthetic, from the back of the hip.

Bone marrow transplant

See stem cell transplant.

Burkitt's lymphoma

A rare, rapidly growing type of B-cell lymphoma (non-Hodgkin's lymphoma). Burkitt's lymphoma needs to be treated as soon as it is diagnosed.

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 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.

Clone

A population of genetically identical cells arising from a single parent cell. Leukaemia is believed to be a clonal disease, that is, all the leukaemia cells may originate from one abnormal 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.

Consolidation treatment

A course of treatment with anti-cancer drugs given to the patient while in remission with the aim of killing any left over cancer cells, and reducing the chances of the disease returning (relapsing) in the future. Also called post-remission therapy.

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.

Hepatomegaly

Enlargement of the liver.

Hickman catheter

A type of central venous catheter (see above) used for patients undergoing intensive treatment such as bone marrow or peripheral blood cell transplantation. It may have a single, double or triple tube (or lumen).

High-dose therapy

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

Immune system

The body's defense 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.

Induction therapy

Treatment given to induce a remission from disease. Induction therapy is the first step in the treatment of acute leukaemias. The aim of this treatment is to destroy any detectable leukaemic cells in the blood and bone marrow and allow the bone marrow to function normally again. Following induction therapy more treatment is given to eliminate any left-over disease in the body.

Intensification

Increasing the amount, number or combination of anti-cancer drugs given to a patient in an attempt to kill drug-resistant or left-over cancer cells in the body.

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.

Maintenance therapy

Treatment given for a period of months or years to maintain a remission and help prevent disease from reappearing (relapsing) in the future. Maintenance therapy is commonly given in the treatment of acute lymphoblastic leukaemia.

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.

Myelo-ablative 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.

Pathologist

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

PICC line

Peripherally inserted central venous catheter (see central venous catheter) inserted in the middle of the forearm.

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 – this is known as hypersplenism.

Splenomegaly

Another term used to describe an enlarged spleen.

Standard therapy

The most effective and safest therapy currently being used.

Stem cells

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





Leukaemia
Foundation

VISION TO CURE
MISSION TO CARE

A bequest

Your planned gift to the Leukaemia Foundation

A wonderful way to make a significant gift is through a bequest in your will. After making due allowance for loved ones, a bequest of a specific amount or a proportion of the residue of your estate, is a way of leaving a real and lasting legacy to the future.

Your bequest to the Leukaemia Foundation will be used to support our mission to care for patients, carers and families and help us achieve our vision to find a cure for leukaemias, lymphomas, myeloma and related blood disorders.

Wording your bequest to the Leukaemia Foundation

You may choose to make a general bequest and allow the Leukaemia Foundation to decide how your bequest will be used, or you may prefer to make that decision yourself e.g. direct your bequest to patient support or research. Your legal adviser can provide further information on the different types of bequests, and on the appropriate wording for a bequest.

As a guide, the following wording may be useful:

'I give and bequeath free of all duties (here state the amount/percentage or share/residue or assets to be gifted) to the Leukaemia Foundation of (here insert the address) absolutely -

- for the general charitable purposes of the said Foundation (this is the Leukaemia Foundation's preferred option); or
- for the purpose of patient and family support; or
- for the purpose of research into the cause, cure or treatment of leukaemia, lymphoma, myeloma and related blood disorders

and I direct that a receipt of the proper officer for the time being of the Leukaemia Foundation shall be a good and sufficient discharge to my trustee/s'.

Please see the next page for the response form.



Response Form

- ☐ I have already made a bequest to the Leukaemia Foundation in my will
- ☐ I am considering/it is my intention to make (please circle) a bequest to the Leukaemia Foundation
- ☐ I would like more information about making a bequest and/or where to direct my bequest
- ☐ I would like to speak to the Planned Giving Manager about appropriate recognition for my bequest
- ☐ I would like to receive invitations to functions

Dr/Mr/Mrs/Ms/Miss:

Address:

..... Postcode.....

Telephone: (h).....

(w)

Email:

Please return this form to the:

Planned Giving Manager,
The Leukaemia Foundation,
GPO Box 9954,
in your Capital City
(marked Private & Confidential)

If you are interested in leaving a bequest to the Leukaemia Foundation in your will and you would like further information, without any obligation, in strictest confidence, please contact the Planned Giving Manager in your state on Freecall 1800 620 420.





Leukaemia
Foundation

VISION TO CURE
MISSION TO CARE

Making a donation

The Leukaemia Foundation is the only national not-for-profit organisation dedicated to the care and cure of patients and families living with leukaemias, lymphomas, myeloma and related blood disorders.

You can help by making a donation. Please fill out the form below or visit www.leukaemia.org.au to make your gift online.

Dr/Mr/Mrs/Ms/Miss:

Address:

..... Postcode.....

Telephone: (h).....

(w)

Email:

Please accept my tax deductible donation for \$

My cheque, made payable to the Leukaemia Foundation, is enclosed, or please charge \$..... to my credit card:

☐ Bankcard ☐ Visa ☐ Mastercard ☐ Amex ☐ Diners

_____/_____/_____/_____

Cardholder's name:

Cardholder's signature:

Expiry date:/.....

Please send to:

The Leukaemia Foundation
GPO Box 9954
in your capital city.





Please send me a copy of the following information booklets:

- ☐ Eating well: a practical guide for people living with leukaemias, lymphomas & myeloma
- ☐ Living with Leukaemias, Lymphomas, Myeloma & Related Disorders, Information and Support
- ☐ Understanding Leukaemias, Lymphomas, Myeloma & Related Disorders
- ☐ Understanding Acute Lymphoblastic Leukaemia in Adults
- ☐ Understanding Acute Lymphoblastic Leukaemia in Children
- ☐ Understanding Acute Myeloid Leukaemia
- ☐ Understanding Allogeneic Transplants
- ☐ Understanding Autologous Transplants
- ☐ Understanding Chronic Lymphocytic Leukaemia
- ☐ Understanding Chronic Myeloid Leukaemia
- ☐ Understanding Hodgkin Lymphoma
- ☐ Understanding non-Hodgkin Lymphoma
- ☐ Understanding Myelodysplastic Syndrome
- ☐ Understanding Myeloma
- ☐ Understanding Myeloproliferative Disorders
- ☐ Young Adults with a Blood Cancer

Or information about:

- ☐ The Leukaemia Foundation's Support Services
- ☐ Workplace giving
- ☐ Regular deduction scheme
- ☐ National Fundraising Campaigns
- ☐ Volunteering
- ☐ Receiving the Foundation's newsletters

Name:

Street or Postal Address:

Suburb:

State/Postcode

Email: Tel: (....).....

Please send to:

Leukaemia Foundation, GPO Box 9954, In Your Capital City

or Freecall 1800 620 420

or email: info@leukaemia.org.au

Further information is available on the Leukaemia Foundation's website
www.leukaemia.org.au





NOTES

First published as *Coping with Childhood Leukaemia* in Jan 2000
Revised and reprinted as *Understanding Acute Lymphoblastic Leukaemia (ALL)*
Children in Oct 2007
Major revision and reprinted May 2010



Leukaemia Foundation

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VISION TO CURE
MISSION TO CARE

This information booklet is produced
by the Leukaemia Foundation and is one in a series on blood
cancers and related disorders.

Some booklets are also available in other languages. Copies of this
booklet and the other booklets can be obtained from the
Leukaemia Foundation in your state by contacting us on

Freecall: 1800 620 420
Email: info@leukaemia.org.au
Website: www.leukaemia.org.au

The Leukaemia Foundation is a non-profit organisation that
depends on donations and support from the community.

Please support our work by calling 1800 620 420
or by mailing your donation to:
The Leukaemia Foundation
GPO Box 9954
in your capital city

May 2010