Acute Lymphoblastic Leukaemia (ALL) in children

A guide for patients and families
# Contents

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acknowledgments</td>
<td>4</td>
</tr>
<tr>
<td>Introduction</td>
<td>5</td>
</tr>
<tr>
<td>The Leukaemia Foundation</td>
<td>6</td>
</tr>
<tr>
<td>Understanding leukaemia</td>
<td>10</td>
</tr>
<tr>
<td>The lymphatic system</td>
<td>16</td>
</tr>
<tr>
<td>What is leukaemia?</td>
<td>18</td>
</tr>
<tr>
<td>What is acute lymphoblastic leukaemia (ALL)</td>
<td>20</td>
</tr>
<tr>
<td>How common is it and who gets it?</td>
<td>21</td>
</tr>
<tr>
<td>Causes</td>
<td>21</td>
</tr>
<tr>
<td>Symptoms</td>
<td>24</td>
</tr>
<tr>
<td>Diagnosis</td>
<td>26</td>
</tr>
<tr>
<td>Types of ALL</td>
<td>30</td>
</tr>
<tr>
<td>Treatment</td>
<td>32</td>
</tr>
<tr>
<td>Commonly used prognostic terms</td>
<td>37</td>
</tr>
<tr>
<td>Fertility</td>
<td>50</td>
</tr>
<tr>
<td>Complementary therapies</td>
<td>52</td>
</tr>
<tr>
<td>Nutrition</td>
<td>52</td>
</tr>
<tr>
<td>Exercise</td>
<td>52</td>
</tr>
<tr>
<td>Making treatment decisions</td>
<td>53</td>
</tr>
<tr>
<td>Social and emotional issues</td>
<td>54</td>
</tr>
<tr>
<td>Useful internet addresses</td>
<td>62</td>
</tr>
</tbody>
</table>
Acknowledgments

The Leukaemia Foundation gratefully acknowledges the following groups who have assisted in the development and revision of the information - people who have experienced childhood leukaemia as a child, parent or carer, Leukaemia Foundation support services staff, nursing staff, and clinical haematologists representing the various states and territories of Australia.

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 Head of Blood Cancer Support at info@leukaemia.org.au.

July 2015
Introduction

This booklet has been written to help parents and families understand more about acute lymphoblastic leukaemia (ALL) in children. The Leukaemia Foundation also has a booklet called ‘Joe has Leukaemia’ which was developed to explain leukaemia to 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 with 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 support services at no charge 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.

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 helped to establish the ALLG Discovery Centre 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 clinicians and allied health professionals.
Foundation staff are health professionals who provide patients and their families with information and support across Australia.

Support Services

The Leukaemia Foundation has a team of highly trained and caring Blood Cancer Support staff with qualifications and experience in nursing and who work across the country.

They can offer individual support and care to you and your family when it is needed.

Support Services may include:

Information

The Leukaemia Foundation has a range of booklets, fact sheets, DVDs 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 our website.

Education and 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 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. Staff may also be able to connect you with others who have experienced a similar diagnosis and treatment regimen.
**Blood buddies**

This is a program for people newly diagnosed with ALL to be introduced to a trained ‘buddy’ who has been living with ALL for at least two years, to share their experience, their learning, and to provide some support.

**Transport**

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

**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. When our accommodation facilities are full, the Foundation may be able to subsidise external accommodation.

*With the cost of hospital car parking and how difficult it can be to find a car park, the Foundation’s transport service has made my hospital visits so much easier.*
Practical assistance
The urgency and lengthy duration of medical treatment can affect everyday life for you and your family and there may be practical things the Foundation can do to help. In special circumstances, the Leukaemia Foundation provides financial support for people who are experiencing financial difficulties or hardships as a result of their illness or its treatment. This assistance is assessed on an individual basis.

Advocacy
The Leukaemia Foundation is a source of support for you as you navigate the health system. While we do not provide treatment recommendations, we can support you while you weigh up your options. We may also provide information on other options such as special drug access programs, and available clinical trials.

Contacting us
The Leukaemia Foundation provides services and support in every Australian state and territory. Every person’s experience of living with ALL is different. Living with ALL is not always easy, but you don’t have to do it alone. Please call 1800 620 420 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
Understanding leukaemia

Leukaemia is the general name given to a group of cancers that develop in the bone marrow. To understand this, we must first understand the role of the bone marrow, stem cells and blood.
Getting to know your bone marrow, stem cells and blood

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 haematopoiesis. There are three main types of blood cells; red cells, white cells and platelets.

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

You might like to think of the bone marrow as the blood cell factory. The main workers at the factory are the stem cells. They are relatively small in number but are able, when stimulated, to reproduce vital 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 the various types of blood cells.

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

Lymphoid (‘lim-foi’d’) stem cells develop into other types of white cells including T-cells, B-cells and Natural Killer Cells.

Red Blood Cells
Carry oxygen for the body to produce energy

White Blood Cells
Form part of the immune system

Platelets
Support blood clotting to stop bleeding
Growth factors and cytokines

All normal blood cells have a limited lifespan in the circulation and need to be replaced on a continual basis. This means that the bone marrow remains very active throughout life. Natural chemicals circulating in your blood called growth factors, or cytokines, control this process of blood cell formation. Each of the different blood cells is produced from stem cells under the guidance of a different growth factor.

Some of the growth factors can now 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 certain white cells, including neutrophils, while erythropoietin (EPO) stimulates the production of red cells.

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.
Blood cells

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. The body uses this oxygen to create energy.

Haematocrit
About 99 per cent of all blood cells in circulation are red blood cells. The percentage of the blood that is occupied by red blood cells is called the haematocrit. A low haematocrit suggests that the number of red cells in the blood is lower than normal.

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

Normal ranges for children:

<table>
<thead>
<tr>
<th></th>
<th>1 month</th>
<th>1 year</th>
<th>3 years</th>
<th>5 years</th>
<th>9 years</th>
<th>16 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemoglobin g/L</td>
<td>102-130</td>
<td>104-132</td>
<td>107-136</td>
<td>110-139</td>
<td>113-143</td>
<td>115-165</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(f) 130-180 (m)</td>
</tr>
<tr>
<td>White cell count x10^{12}/L</td>
<td>6.4-12.1</td>
<td>5.4-13.6</td>
<td>4.9-12.8</td>
<td>4.7-12.3</td>
<td>4.7-12.2</td>
<td>3.5-11</td>
</tr>
<tr>
<td>Platelets x10^{12}/L</td>
<td>270-645</td>
<td>205-553</td>
<td>214-483</td>
<td>205-457</td>
<td>187-415</td>
<td>150-450</td>
</tr>
<tr>
<td>Neutrophils</td>
<td>0.8-4.9</td>
<td>1.1-6.0</td>
<td>1.7-6.7</td>
<td>1.8-7.7</td>
<td>1.8-7.6</td>
<td>1.7-7.0</td>
</tr>
</tbody>
</table>
White cells

White cells, also known as leukocytes, fight infection. The following is a list of some of the different types of white cells:

**Neutrophils:** mainly kill bacteria and remove damaged tissue. Neutrophils are often called the first line of defence when infections occur. They are often the first white blood cell at the site of infection and attempt to destroy the foreign pathogen before it becomes a problem to the body.

**Eosinophils:** mainly kill parasites

**Basophils:** mainly work with neutrophils to fight infection

**Monocytes:** mainly work with neutrophils and lymphocytes to fight infection; they also act as scavengers to remove dead tissue. These cells are known as monocytes when found in the blood, and called macrophages when they migrate into body tissue to help fight infection.

**B-cells:** mainly make antibodies that target micro-organisms, particularly bacteria.

**T-cells:** mainly kill viruses, parasites and cancer cells and produce cytokines which can recruit other cells to make antibodies which target micro-organisms.

These white cells work together to fight infection as well as having unique individual roles in the fight against infection.

**Neutropenia**

Neutropenia is the term given to describe a lower than normal neutrophil count. If you have a neutrophil count of less than $1 \times 10^9/L$, you are at an increased risk of developing more frequent and sometimes severe infections.

---

To help prevent infection, regular washing of my hands has become part of my new normal.
**Platelets**

Platelets are cellular fragments that circulate in the blood and play an important role in clot formation. They help to prevent bleeding.

If a blood vessel is damaged (for example by a cut) the platelets gather at the site of the injury, stick together and form a plug to help stop the bleeding. They also release chemicals, called clotting factors, that are required for the formation of blood clots.

**Thrombocytopenia**

Thrombocytopenia is the term used to describe a reduction in the platelet count to below normal. If your platelet count drops too low, you are at an increased risk of bleeding and tend to bruise easily. Each treatment centre will have their own guidelines on the specific platelet count level when interventions may need to be taken. Platelet transfusions are sometimes given to return the platelet count to a safer 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.

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 the stomach, gut and skin.
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 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.
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 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.
What are the different 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 12).

**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 12).

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.
Acute lymphoblastic leukaemia (ALL)

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

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

**How common is it and who gets it?**

Each year in Australia around 250 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.
Causes

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.

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 on the next page.

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 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. 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 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 syndrome and Fanconi anaemia are at an increased risk of developing ALL.
**Symptoms**

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

**Anaemia**

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

**Increased bleeding or bruising**

A very low platelet count can cause bruising for no apparent reason, or excessive or prolonged bleeding following minor cuts or injury.

Some 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 ('pet-tie-kee-ə') 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. 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 the 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.
Diagnosis

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), full blood examination (FBE), 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), a low platelet count and a low neutrophil count. This is sometimes called pancytopaenia. Most children will have abnormal leukaemic blast (immature) cells in their bloodstream.

Depending on how many leukaemic blasts have spilled out of the bone marrow and into the bloodstream, the total white blood cell count can be elevated, normal or low. 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 per cent and almost 100 per cent in children with ALL.

The bone marrow examination will be done in the hospital. All children should 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 usually collected from the back of the hip bone, 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 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, cytogenetic, and MRD 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. The test is performed using a technique called flow cytometry or flow, for short.

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, rearranged 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 blast cells. They are not passed down from parent to child (inherited).

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. Certain cytogenetic abnormalities are associated with better outcomes, so are called favourable cytogenetics.

Other abnormalities are associated with an increased risk of relapse, so are often referred to as high risk or unfavourable cytogenetics. Thankfully, high risk cytogenetic features are relatively uncommon in childhood ALL.
I seemed to be having so many tests. I just wanted to know what was wrong inside my body. The waiting and the unknown were the worst part for me.

The results of cytogenetic testing can take a week or two to come back. How the cytogenetic results influence your child’s management usually depends on both the specific abnormalities present in the leukaemic blasts and your child’s response to their initial treatment. Your doctor will discuss the results and their impact on treatment with you.

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 (LP), which is usually performed under a short general anaesthetic or local anaesthetic and usually at the same time as the first bone marrow examination.

If there is enough concern that your child has leukaemia, a single dose of intrathecal (IT) chemotherapy will usually be given into the CSF at the same time as this first lumbar puncture.

The CSF fluid is then tested in the laboratory to check for the presence of leukaemia 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, heart 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. An ultrasound, or echocardiogram (“echo”) of the heart is also usually done to check heart function.

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.
Types of ALL

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
- ALL that arises in developing T-cells

The World Health Organisation’s classification system for ALL uses information obtained from looking at the leukaemia under the microscope, together with 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. Subtypes of ALL depends on the presence or absence of distinct cell surface markers.

B precursor (Pre-B-cell) ALL

In around 80 per cent 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 or simply B-ALL.

Mature B-cell ALL

Mature B-cell ALL arises in more mature developing lymphocytes. This type of ALL is less common accounting for around five per cent 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. Mature B-cell ALL is biologically similar to another disease called Burkitt’s lymphoma, a rare aggressive type of non-Hodgkin lymphoma. Children diagnosed with mature B-cell ALL are treated with similar drugs to those used to treat Burkitt’s lymphoma and it is sometimes referred to as Burkitt’s leukaemia.
T-cell ALL

In around 15 per cent of cases ALL arises in developing T-cells in the thymus gland in the chest and is called T-ALL. Children with T-ALL often have a high white blood cell count and involvement of the central nervous system at diagnosis.

In around 50 per cent of cases, the thymus gland is enlarged and visible on x-rays in the centre of the chest (mediastinal mass). In some cases there may also be fluid in the chest around the lungs, called a pleural effusion. If the mediastinal mass or pleural effusion is very large or the child is having breathing difficulty, a general anaesthetic for a diagnostic bone marrow aspirate and lumbar puncture may not be safe.

The diagnosis of T-ALL may then need to be made by draining some of the fluid from the chest under sedation and analysing that fluid in the laboratory. Usually there are enough leukaemic blasts in the pleural effusion to make the diagnosis. Treatment is then started to shrink down the mediastinal mass, allowing a bone marrow aspirate and lumbar puncture to be done under general anaesthetic safely.
Treatment

**ALL usually progresses quite quickly so treatment needs to begin as soon as possible.**

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 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) aimed at improving the way this disease is 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 word “risk” refers to the risk of relapse. 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.

Risk groups are usually referred to as standard (or average), low (or favourable), high or very high. 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 may give people access to new therapies that are still undergoing research and not yet funded by governments.

Although new drugs or treatment approaches are expected to give better outcomes, they usually need to be compared to the current best standard treatment (the ‘standard arm’) in a randomised way to avoid any potential bias and confirm they improve outcomes. If enrolled on a randomised trial, patients are allocated randomly between the ‘standard arm’ of the study and the ‘randomised arm’ with the new drug or treatment approach.

As parents you will need to give your informed consent 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.

If you do not agree to be enrolled on a clinical trial, you will still receive the current best-practice treatment, care and support at your treating centre.

Your child’s haematologist will treat your child according to the ‘standard arm’ of the clinical trial, as this usually represents the current best treatment for your child’s disease. This is often referred to being treated ‘off study’. 
My treatment started so quickly after my diagnosis. I was so scared but thankful that they were treating me straight away. I wanted to get on top of this thing inside of me.

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 or leukaemia 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), or 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 the central nervous system (CNS).

Intravenous drugs are usually given through a special line called a central venous catheter (or central line) or a port-a-cath (or port, or infusaport). These are special lines inserted surgically 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 five per cent. 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 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 testes, 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 delayed intensification)
• Maintenance therapy

Depending on the protocol your child is following, remission induction, consolidation, interim maintenance and delayed 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.
It was quite hard being diagnosed as a young person. I couldn't do the things I enjoyed with my friends.

Consolidation therapy

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 and includes interim maintenance and delayed 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 treatment with varying intensity over several months and includes additional drugs such as 6-mercaptopurine, methotrexate, cyclophosphamide, cytarabine, etoposide and thioguanine. This is given to reduce left over disease that may be present, even if it can’t be detected with tests.

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 (also called Philadelphia positive ALL or Ph+ ALL) has been associated with a poorer prognosis using standard therapy.

Taking these and other factors into consideration, children are categorised as having low, standard, high or very 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 (15-20 per cent) will experience a relapse over time.

**Minimal residual disease (MRD)**

We now know that there is usually a strong relationship between the number of leukaemic cells left over in a child’s body following initial 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 two years. Depending on the type of ALL and the protocol, boys often receive a third year of maintenance therapy. 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 six months after your child has finished treatment. If your child has a stem cell transplant, immunisations may be delayed for six to 12 months afterwards. Certain vaccinations, such as the seasonal influenza vaccine, may be given during treatment.
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.

**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.

*The Leukaemia Foundation has produced books for children to understand stem cell transplants*
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 (usually defined as being less than $1 \times 10^9/L$, or simply ‘less than 1’).
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:

- two temperatures between 38ºc and 38.4ºc within an hour, or a single temperature of 38.5ºc or higher (even if it returns to normal) and/or an episode of uncontrolled shivering (a rigor)

- bleeding (or bruising), for example blood in the urine, bowel motions, coughing up blood, bleeding gums or a persistent nose bleed

- prolonged nausea or vomiting that prevents them from eating or drinking or taking their normal medications

- diarrhoea, stomach cramps, abdominal pain or severe constipation

- back pain

- 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 bandannas. 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, many, 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.

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 (e.g. 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 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 topped up with some intravenous fluid in the hospital day treatment centre or be admitted to hospital.

Appetite
Some children may lose their appetite during more intensive phases of chemotherapy. However, many more children put on weight during treatment and obesity is a common problem in survivors in later life. It is important to continue normal healthy eating patterns in the family.

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 (e.g. chips and chocolate), and high-sugar 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, aggression, sadness, 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.

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.

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 co-trimoxazole which is used to help prevent an infection called Pneumocystis carinii pneumonia or PCP (now officially called Pneumocystis jiroveci). This is an organism that people are commonly exposed to but can cause severe pneumonia when the immune system is compromised (such as patients on chemotherapy).

Treatment with co-trimoxazole 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, and 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 young or 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 such things as herbs and vitamins, 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 while no complementary or alternative treatment alone has proven to be effective against childhood ALL, some complementary therapies may help improve the child’s quality of life, and when utilised in conjunction with the treating team, may reduce treatment-related side effects.

It is 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, nurse, or dietitian 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 the dietitian about 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.

Exercise

Exercise is being utilised more often in wellness centres, and treatment centres to help to maintain the child’s physical development, maintain fine motor skills, reduce fatigue and boredom, and improve treatment recovery times.

Speak with your treatment centre about appropriate exercise for your child. You may wish to request a referral to an exercise physiologist for guidance.
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.

After a diagnosis, the whole family can be affected in unexpected ways.

Relationships may be altered – either strengthened, or damaged due to the different coping strategies people utilise to get through a very difficult time.

Siblings may feel left out and act out to get attention. Change in the family dynamic is expected at this time, and people should seek support from the treatment team to ensure the family is maintained during and after this treatment period.

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 having their port-a-cath accessed or other bloods tests, 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.

Your treatment team can help support you in communicating important information to your child in an age/development appropriate way.

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 (six to 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.

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, 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 Ronald McDonald House, CanTeen, Camp Quality, 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 may experience many confusing emotions. The way 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.

Support may also be available to siblings through the Leukaemia Foundation, CanTeen and Camp Quality.

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.

After treatment, a care plan should be developed by your child’s treating team outlining the expected future care, anticipated appointment time frames, what precautions should be taken (e.g. increased screening and tests for potential complications), what lifestyle adjustments should be made (e.g. exercise or dietary needs) and when/if
you should contact the specialist. Both you and your GP should be given a copy of this document. It is sometimes called a survivorship care plan.

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 parents 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 it 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

Leukaemia Foundation of Queensland
www.leukaemiaqld.org.au

American Cancer Society
www.cancer.org

Australia and New Zealand Children’s Haematology/Oncology Group (ANZCHOG)
www.ANZCHOG.org

Australian Bone Marrow Donor Registry
www.abmdr.org.au

Blood & Marrow Transplant Information Network (USA)
www.bmtinfonet.org

Bloodwise (UK)
www.bloodwise.org.uk

Camp Quality
www.campquality.org.au

Cancer Council of Australia
www.cancercouncil.com.au

CanTeen
www.canteen.org.au

CureSearch
www.curesearch.org

Leukemia & Lymphoma Society (USA)
www.lls.org

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/about-cancer

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

**Antibodies**
Naturally produced substances in the blood, made by white blood cells called B-lymphocytes or B-cells. Antibodies target antigens on 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.

**Anti-viral**
A drug used to prevent or treat specific viral infections such as cold sores or chicken pox.
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 centre of many flat or big bones of the body. Active or red bone marrow contains stem cells from which all blood cells are made and in the adult this is found mainly in the bones making up the axial skeleton – hips, ribs, spine, skull and breastbone (sternum).

The other bones contain inactive or (yellow) fatty marrow, which, as its name suggests, consists mostly of fat cells.

Bone marrow 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), central line or Hickman line. A tube passes through the large veins of the neck, chest or groin and into the central blood circulation. It can be used for taking samples of blood, giving intravenous fluids, blood, chemotherapy and other drugs without the need for repeated needles.

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

Chemotherapy
Single drugs or combinations of drugs which may be used to kill and prevent the growth and division of cancer cells. Although aimed at cancer cells, chemotherapy can also affect rapidly dividing normal cells and this is responsible for some common side-effects including hair loss and a sore mouth. Nausea and vomiting are also common, but nowadays largely preventable with modern anti-nausea medication. Most side-effects of are temporary and reversible.
**Chromosomes**
Chromosomes are made up of coils of DNA (deoxyribonucleic acid). DNA carries all the genetic information for the body in sequences known as genes. There are approximately 40,000 genes on 23 different chromosomes. The chromosomes are contained within the nucleus of a cell.

**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 readily detected. In people with leukaemia this means that the proportion of blast cells in the marrow has been reduced to less than 5 per cent. There are no blast cells present in the circulating blood 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 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 leftover cancer cells, and reducing the chances of the disease returning (relapsing) in the future. Also called post-remission therapy.

**Corticosteroids (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, corticosteroids 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 centre 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).

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

**Haemopoiesis**
The formation of blood cells.

**Hepatomegaly**
Enlargement of the liver.

**Hickman catheter**
A type of central venous catheter (see above) used for patients undergoing intensive treatment for high-risk leukaemia and/or haemopoietic stem 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 defence system against infection and disease.

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

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

**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, which are 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 ALL and involves outpatient-based oral chemotherapy with or without intermittent IV and IT chemotherapy.

**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. Spontaneous mutations occur commonly in normal cells, but normal cells have the ability to either repair the mutation or undergo programmed cell death. Cancer cells have usually lost the ability to repair such mutations and continue to grow and divide.

**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.
Neutropenia
A reduction in the number of circulating neutrophils, an important type of white cell. Neutropenia 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.

Port-a-cath (infusaport)
A central venous catheter inserted surgically, ending in a round plastic ‘hub’ that sits under the skin. Also referred to as ‘ports’ or sometimes by children as ‘buttons’. Special L-shaped needles are inserted through the skin to ‘access’ the port-a-cath when needed for giving fluids, chemotherapy or other drugs and can also be used for taking blood tests.

Prognosis
An estimate of the likely course of a disease.

Protocol
A treatment program detailing which chemotherapy drugs are given during different stages of treatment. Clinical trial protocols usually compare a new drug or treatment approach (the ‘randomised arm’) to the current best standard treatment (the ‘standard arm’).

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 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.
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 leukaemia, lymphoma, myeloma and related blood disorders. The Foundation receives no ongoing government support and relies on the generosity of the community to support our Vision to Cure and Mission to Care.

How can I give?

ONLINE www.leukaemia.org.au

PHONE 1800 620 420

POST (complete this form or enclose cheque/money order and return)
The Leukaemia Foundation, Reply Paid 9954 in your capital city

Name

Address

Postcode

Phone

Mobile

Email

I enclose my gift of (please tick box)

$30  $50  $75  $100  $250  Other $

I wish to make a regular monthly donation of $  Commencing on  /  /  *

*You can cancel at any time by calling 1800 620 420.

My cheque/money order made payable to the Leukaemia Foundation is enclosed.

I wish to pay with my credit card and my details are included below:

Visa  MasterCard  Diners  Amex

Card Number

Expire Date  /  CVV

Cardholder’s Name

Signature

Your privacy is important to us. That is why we treat your personal information with confidence. To learn more about how and why we collect and use any personal or sensitive information about you, please view our Notification Statement at www.leukaemia.org.au/privacy
Please send me a copy of the following booklets:

- Leukaemia, Lymphoma, Myeloma, MDS, MPN and related blood disorders
- Acute Lymphoblastic Leukaemia in Adults (ALL)
- Acute Lymphoblastic Leukaemia in Children (ALL)
- Acute Myeloid Leukaemia (AML)
- Amyloidosis
- Chronic Lymphocytic Leukaemia (CLL)
- Chronic Myeloid Leukaemia (CML)
- Hodgkin Lymphoma
- Non-Hodgkin Lymphoma (NHL)
- Myelodysplastic Syndrome (MDS)
- Myeloma
- Myeloproliferative Neoplasms (MPN)
- Eating Well
- Living with Leukaemia, Lymphoma, Myeloma, MDS, MPN and related blood disorders
- Autologous Stem Cell Transplants (also called Bone Marrow Transplants)
- Young Adults with a Blood Cancer
- My Haematology Diary
- Tom has Lymphoma
- Joe has Leukaemia
- Ben’s Stem Cell Transplant
- Jess’ Stem Cell Donation

Books for children:

- The Leukaemia Foundation’s Support Services
- Giving at work
- Monthly giving program
- National fundraising campaigns
- Volunteering
- Receiving our newsletters
- Leaving a gift in my will

Or information about:

Name
Address
Postcode
Phone
Mobile
Email

POST TO The Leukaemia Foundation, Reply Paid 9954 in your capital city
PHONE 1800 620 420 EMAIL info@leukaemia.org.au
FURTHER INFORMATION ONLINE www.leukaemia.org.au
This information booklet is produced by the Leukaemia Foundation and is one in a series on leukaemia, lymphoma, myeloma, MDS, MPN and related blood disorders.

Copies of this booklet can be obtained from the Leukaemia Foundation in your state by contacting us.

The Leukaemia Foundation is a not-for-profit organisation that depends on donations and support from the community. Please support our work.

July 2015

Contact us

1800 620 420

GPO Box 9954, IN YOUR CAPITAL CITY

info@leukaemia.org.au

leukaemia.org.au