

UNDERSTANDING MYELODYSPLASTIC SYNDROME (MDS)

A guide for people with MDS
and their families



Leukaemia
Foundation

VISION TO CURE
MISSION TO CARE

CONTENTS

	PAGE
Introduction	3
The Leukaemia Foundation	4
Bone marrow stem cells and blood cell formation	8
What is MDS?	13
Who gets MDS?	14
How common is MDS?	14
What causes MDS?	14
What are the symptoms of MDS?	16
How is MDS diagnosed?	17
Types of MDS	20
Prognosis	22
Treatment for MDS	23
Making treatment decisions	38
Information and support	39
Useful internet addresses	40
Glossary of terms	41

ACKNOWLEDGEMENTS

The Leukaemia Foundation gratefully acknowledges the following groups that have assisted in the development and revision of the information - people who have experienced MDS personally or as a carer, Leukaemia Foundation support services staff, nursing staff, and clinical haematologists representing the various states and territories of Australia. The cartoon illustrations were drawn by Brett Hansen.

The Leukaemia Foundation values feedback from people who have been affected by MDS and their loved ones, and health care professionals working with people with MDS. If you would like to make suggestions, or tell us about your experience of using this booklet, please contact the Head of Support Services at

info@leukaemia.org.au.

The Leukaemia Foundation also gratefully acknowledges Novartis for their support in the production of this booklet through an unrestricted educational grant.



Leukaemia
Foundation

VISION TO CURE
MISSION TO CARE

INTRODUCTION

This booklet has been written to help you and your family understand more about myelodysplastic syndrome (MDS), also known as myelodysplasia.

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

You may not feel like reading this booklet from cover to cover. It might be more useful to look at the list of contents and read the parts that you think will be of most use at a particular point in time.

We have used some medical words and terms 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.

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

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

THE LEUKAEMIA FOUNDATION

The Leukaemia Foundation is Australia's peak body for blood cancer in Australia, funding research and providing free services to support people with leukaemia, lymphoma, myeloma, MDS, MPNs and related disorders. Since 1975, the Foundation has been committed to improving survival for people diagnosed and providing much needed support. It does not receive direct ongoing government funding, relying instead on the continued and generous support of individuals and corporate supporters to develop and expand its services.

Our support may be offered over the telephone, face to face at home, hospital or at the Foundation's accommodation centres, depending on geographical and individual needs. Support may include providing information, education seminars and programs that provide a forum for peer support and consumer representation, practical assistance, accommodation, transport, emotional support, and advocacy.

The Leukaemia Foundation funds leading research into better treatments and cures for blood cancers and related blood disorders. Through its National Research Program, the Foundation has established the Leukaemia and Lymphoma Tissue Bank at the Princess Alexandra Hospital and the Leukaemia Foundation Research Laboratory at the Queensland Institute for Medical Research. In addition, the Foundation funds research grants, scholarships and fellowships for talented researchers.

Support Services



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

The Leukaemia Foundation has a team of highly trained and caring Support Services staff with qualifications in nursing or allied health in every State and Territory across the country. We 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 the website.

Education & support programs

The Leukaemia Foundation offers you and your family MDS-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 MDS can have a dramatic impact on a person's life. At times it can be difficult to cope with the emotional stress involved. The Leukaemia Foundation's Support Services staff can provide you and your family with much needed support during this time. They may refer you or a loved one to a specialist health professional eg psychologist if required

Online discussion forum

The Foundation has established an on-line information and support group for people living with a blood cancer or related blood disorder. A section specific to MDS is available for people to talk about their experiences, ask questions of others with the disorder to learn from their experience, and to connect with others who understand what it is like to live with MDS. Registration is free and participants can remain anonymous, see www.talkbloodcancer.com

Telephone discussion forums

This support service enables anyone throughout Australia who has MDS to share their experiences, provide tips, education and support to others in a relaxed forum. Each discussion forum is facilitated by a member of the Leukaemia Foundation Support Services Team who has a background in haematology nursing.

Accommodation

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

Transport

The Foundation also assists with transporting people to and from hospital for treatment or follow up appointments relating to their blood cancer. Courtesy cars and other services are available in many areas throughout the country.

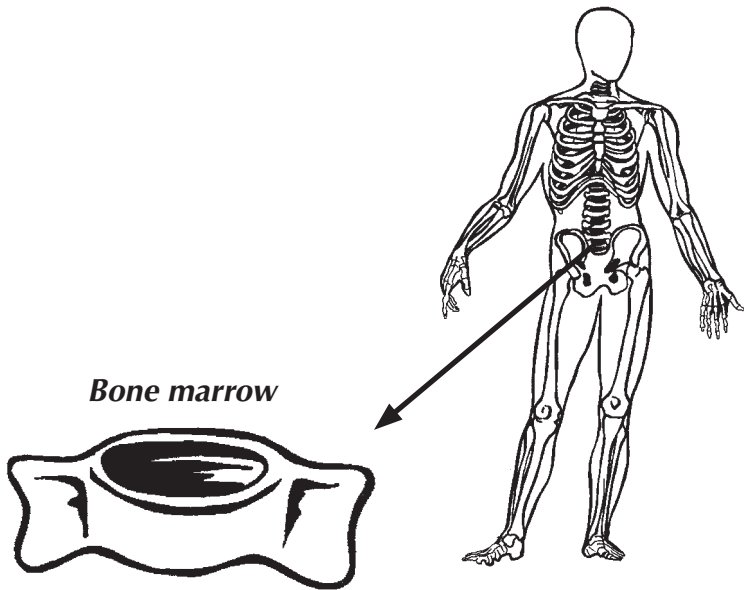
Practical assistance

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

Contacting us

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

BONE MARROW, STEM CELLS AND BLOOD CELL FORMATION



Bone marrow

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

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

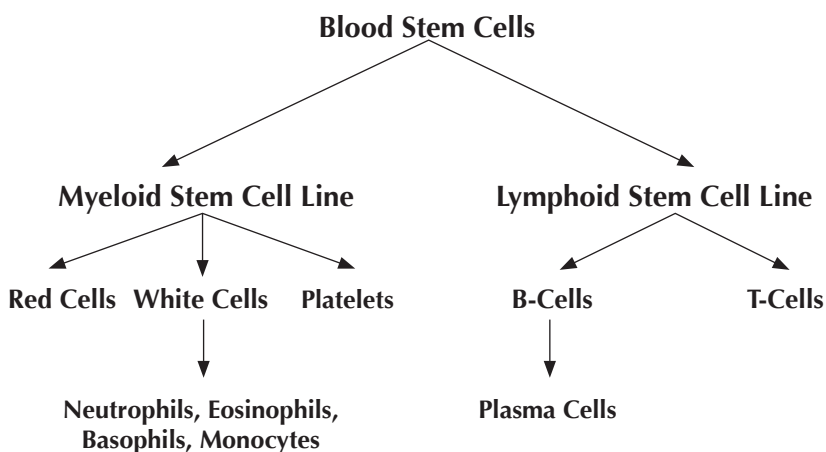
You might like to think of the bone marrow as the blood cell factory. The main workers at the factory are the blood *stem cells*. They are relatively small in number but are able, when stimulated, to reproduce vital numbers of red cells, white cells and platelets. All blood cells need to be replaced as they have a limited life span.

There are two main families of stem cells, which develop into various types of blood cells.

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

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

Growth factors and cytokines



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

These days some growth factors can be made in the laboratory (synthesised) and are available for use in people with blood disorders. For example, *granulocyte colony stimulating factor* (GCSF) stimulates the production of white cells called *neutrophils* while erythropoietin (EPO) stimulates the production of red cells.

Blood

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

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.

Fast Fact: Did you know that on average, a person will produce around two million red cells every second?



Plasma

60%

Blood cells

40%

The normal haemoglobin (Hb) range for a man is approximately 130 - 170 g/L

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

Haematocrit

Red cells are by far the most numerous blood cell and 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.

The normal range of the haematocrit (HCT) in a man is between 40% and 52%

The normal range of the haematocrit in a woman is between 36% and 46%

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

Low Hb=low oxygen=low energy

White cells

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

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

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

The normal adult white cell count varies between 3.7 and 11 x 10⁹/L

Neutropenia

Neutropenia is the term used to describe a lower than normal neutrophil count. If you have a neutrophil count of less than 1 (1 x 10⁹/L) you are considered to be neutropenic and at risk of developing frequent and sometimes severe infections.

The normal adult neutrophil count varies between 2.0 and 7.5 x 10⁹/L

Platelets

Platelets are fragments of a cell called a megakaryocyte. They circulate in the blood and play an important role in blood clot formation. They help to prevent bleeding. If a blood vessel is damaged (e.g. by a cut) the platelets gather at the site of injury, stick together to form a plug to help stop the bleeding. They also release chemicals required for blood clot formation.

*The normal adult platelet count
varies between 150 and 400 x 10⁹/L*

Thrombocytopenia

Thrombocytopenia is the term used to describe a reduction in the platelet count to below normal. If your platelet count drops below 20 (20 x 10⁹/L) you are at a higher risk of bleeding and tend to bruise easily. Some people are at a higher risk of bleeding than others, and platelet transfusions are sometimes given to bring the platelet count back to a safer level.

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

WHAT IS MDS?

Myelodysplastic syndrome (MDS) is a group of diseases which all affect, to a greater or lesser extent, the production of normal blood cells in the bone marrow.

In MDS, abnormal bone marrow stem cells produce increased numbers of immature blood cells (called blast cells). These cells do not grow properly and often die prematurely. This results in lower numbers of normal mature red blood cells, white blood cells and platelets being produced. The blood cells that do survive are often of poor quality, are abnormal in shape and appearance (dysplastic) and unable to function properly.

The release of these abnormal cells from the bone marrow into the blood stream is also defective. This means that people with MDS often have a very active bone marrow but a low number of functional blood cells circulating in the blood stream. Without enough red blood cells, white blood cells and platelets you can become fatigued, more susceptible to infections, and may bleed and bruise more easily.

In approximately 15 per cent of cases, people with MDS have very low numbers of cells in their bone marrow. This is referred to as 'hypoplastic myelodysplasia'.

There are different types of MDS and the disease can vary in its severity and the degree to which normal blood cell production is affected. People with mild disease are often found to simply be anaemic, or they might have a lower than normal white blood cell or platelet count, but in many cases they have few, if any, troubling symptoms from their disease. In more severe cases, the lack of circulating blood cells is more pronounced, causing more symptoms.

Some cases of MDS, approximately 30 per cent, have the potential to progress to acute myeloid leukaemia (AML) and MDS is therefore often referred to as a pre-leukaemic disease.

WHO GETS MDS?

Over 90 per cent of cases occur in people over the age of 60, but MDS can occur at any age, including very occasionally in children.

HOW COMMON IS IT?

Overall, MDS is relatively uncommon, with around 1300 Australians diagnosed each year. It is commonly considered a haematological or blood disorder of the elderly.

It's difficult to be sure of the exact number of people who have MDS. This is because in many cases the disease develops slowly and people don't have any symptoms for a long time. In these cases MDS may go undetected for several years.

MDS is a notifiable disease, and a diagnosis needs to be recorded on Australian cancer registries. Reports on the national cancer registry are provided by the Australian Institute of Health and Welfare: www.aihw.gov.au.

WHAT CAUSES MDS?

MDS occurs as a result of a mutation (a fault or change) in one or more of the genes that control blood cell development. This change (or changes) results in the abnormal growth of blood stem cells. The original mutation is preserved when the affected stem cell divides and produces a 'clone'; that is a group of identical cells all with the same defect. This is why MDS is sometimes described as a 'clonal blood stem cell disorder', and is a "cancer-like" disease.

Mutations in dividing cells occur all the time, and cells have clever ways of stopping these abnormalities persisting and causing problems within the body. The longer we live, however, the more chance we have of acquiring mutations that manage to escape these safe-guards. That is why MDS, like most leukaemias and other cancers, becomes more common as we get older. This naturally occurring or spontaneously-arising MDS is referred to as primary MDS. It is not contagious; you cannot 'catch' it by being in contact with someone who has the disease and it is not inherited or passed on within families.

Why MDS occurs – some known risk factors:

Any process which damages genes and leads to mutations may have a role in the development of MDS.

Ageing

Ageing appears to be the most important risk factor for MDS because the risk of developing mutations increases with age.

Chemicals

Exposure to high levels of some environmental chemicals, especially benzene and petroleum products, is associated with the development of MDS.

Cigarette smoking

Exposure to chemicals in tobacco smoke may increase the risk of developing MDS. .

Cytotoxic chemotherapy

People previously treated for cancer or other conditions with chemotherapy, are at an increased risk of developing what is called secondary or treatment-related MDS. This accounts for less than 10 per cent of all cases of MDS. Secondary MDS is associated with different mutations than those that occur in spontaneous MDS, and can be more challenging to treat. The time between exposure to the drugs and development of MDS may be anything from 2-3 years to over 10 years.

Radiation

Previous radiation therapy, or accidental exposure to high levels of environmental irradiation, is associated with an increased risk of MDS. In some cases it may not be apparent for up to 40 years.

Rare congenital or familial causes

Certain congenital disorders such as Bloom's Syndrome, Down's Syndrome, Fanconi Anaemia and neurofibromatosis have unstable genes and are more at risk of developing mutations that cause MDS or cancer.

WHAT ARE THE SYMPTOMS OF MDS?

Many people in the early stages of MDS have no symptoms at all and it may be diagnosed during a routine blood test. In other cases people go to see their general practitioner (GP) because they have some troubling symptoms from their disease. The types of symptoms that people experience depend on how severe their disease is and the type of blood cell which is most affected.

The most common symptoms are caused by a lack of red cells (anaemia):

- persistent tiredness and fatigue
- weakness
- shortness of breath or chest pains with minimal exercise
- looking pale

Abnormal white cell function, often with low white cell counts, causes:

- recurring infections, especially chest infections
- fevers
- sore mouth due to mouth ulcers

Abnormal platelet function, often with low platelet counts, causes:

- easy bruising
- purpura – a rash of small red dots, seen often on the lower limbs initially, due to small surface capillary bleeds which are known as petechiae
- tendency to bleed, especially from the nose and gums

Many people with MDS have a combination of symptoms. This is because the production of all of the blood cell types may be affected by the disease.

HOW IS MDS DIAGNOSED

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

Full blood count

The first step in diagnosing MDS requires a simple blood test called a full blood count (FBC) or complete blood count (CBC). A sample of blood is drawn from a vein in your arm, sent to the laboratory where the blood cells are counted, and the blood is examined under the microscope.

The number of red cells, white cells and platelets, and their size, shape and appearance is noted as these can all be abnormal in MDS. Other blood tests will be done to exclude other causes of anaemia, for example; low iron, folate and vitamin B12 levels and tests of thyroid function to exclude another common cause of fatigue.

Bone marrow biopsy

If the results of your blood tests suggest that you might have MDS, a bone marrow biopsy may be required to help confirm the diagnosis. A bone marrow biopsy involves taking a sample of bone marrow, usually from the back of the hip bone and sending it to the laboratory for examination under the microscope.

The sample of bone marrow is examined in the laboratory to determine the number and type of cells present and the amount of haemopoiesis (blood forming) activity taking place there. Although the blood counts are low in the majority of cases of MDS, the bone marrow is very active but with increased numbers of immature cells that are abnormal in shape, appearance and size. In addition, blood cell production is usually found to be very inefficient; this is referred to as ineffective haematopoiesis. The percentage of blast cells (the abnormal stem cells) seen in the bone marrow (and sometimes in the blood) gives a guide to the severity of the myelodysplasia. The results of this bone marrow test may take a few days to be finalised.

The bone marrow biopsy may be done in the haematologist's rooms, clinic or day procedure centre and is usually performed under local anaesthesia with sedation given either by tablet or through a small drip in your arm. In selected cases a short general anaesthetic may be required. The local anaesthetic is given as an injection under the skin. The injection takes a minute or two, and you should feel only a mild stinging sensation.

After allowing time for the local anaesthetic to work, a needle is inserted through the skin and outer layer of the bone into the bone marrow cavity. A syringe is attached to the end of the needle and a small sample of bone marrow fluid is drawn out - this is called a 'bone marrow aspirate'. Then the 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'.

You might feel a bit drowsy afterwards, so you should take a family member or friend along who can take you home. A small dressing or plaster over the biopsy site can usually be removed the next day. There may be some mild bruising. Mild discomfort, is usually managed affectively by paracetamol. More serious complications such as bleeding or infection are very rare.

You have the option to refuse sedation during the procedure if you have to drive yourself home from the clinic after the procedure and you are concerned about the effects of the sedation.

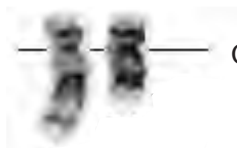
Waiting around for tests can be both stressful and boring. Remember to ask beforehand how long the tests will take and what to expect afterwards. You might like to bring a book, some music or a friend for company and support.

Cytogenetic tests

Cytogenetic tests provide information about the genetic make-up of the 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 genetic abnormalities, such as missing or extra chromosomes, are a clue to the underlying mutation in the DNA of the gene. Not all mutations cause obvious changes to the chromosomes, but abnormalities may be detected in about 60 per cent of cases. These may help to confirm the subtype of MDS you have and provide information about the likely course of your disease (prognosis) and the best way to treat it. One of the most common cytogenetic abnormalities found in MDS is the deletion of part of the long arm of chromosome 5 known as deletion 5q, del(5q) or 5q-. This is illustrated below. It may take a few weeks to obtain a result from these cytogenetic tests.

Normal Chromosome 5



Chromosome del(5q)

TYPES OF MDS

The World Health Organisation's 2008 classification system recognises several major subtypes of MDS. These subtypes are distinguished from each other by the degree to which normal blood cell production is affected, the number of blast cells present and the likelihood of transformation to acute myeloid leukaemia.

Knowing the exact type of MDS you have is important because it helps the doctor to decide on the best course of treatment for you.

Major Subtypes of MDS (based on the World Health Organisation (WHO) Classification)

Refractory cytopenia with unilineage dysplasia (RCUD) and Refractory cytopenia with multilineage dysplasia (RCMD)

"Cytopenia" means a reduction in one or more types of cell in the blood. In the RCUD type of MDS, the red blood cells are most commonly affected, causing anaemia ("Refractory Anaemia" - RA). Sometimes the platelets or neutrophils are the worst affected. If more than one type of cell is affected in the blood and/or bone marrow it is said to be "multilineage" and the diagnosis is RCMD. Most importantly, in RCUD and in RCMD types of MDS, the bone marrow contains less than 5% abnormal blast cells and there are none found in the circulating blood. This type of MDS rarely transforms to leukaemia. Treatment is usually regular observation or blood transfusion only, unless the other blood counts are severely affected.

Myelodysplastic syndrome with isolated del(5q) chromosome

Red blood cells are affected, causing anaemia. There is usually less than 5% blast cells in the bone marrow and circulating blood. The developing blood cells in the bone marrow display the unique chromosome abnormality del(5q), where a part of chromosome 5 is not present. This is an example of a chromosome abnormality associated with a good prognosis.

Refractory anaemia with ring sideroblasts (RARS)

Similar to refractory anaemia, but in this case the red blood cells are unable to process the iron that normally goes into making haemoglobin, the oxygen carrying component of the red cell. Instead the iron granules form a ring around the nucleus of a developing red blood cell. These are called 'ring sideroblasts', and can be seen under the microscope. In most cases the developing cells in the bone marrow contain a mutation in a gene called SF3B1.

Refractory anaemia with excess blasts – type 1 (RAEB-1)

One or more blood cell types are affected. The bone marrow contains between 5% and 9% blast cells (in normal bone marrow there should be less than 5%) and there are only a small number of blast cells (less than 5%) found in the circulating blood (no blast cells are found in normal circulating blood).

Refractory anaemia with excess blasts – type 2 (RAEB-2)

One or more blood cell types are affected, but this time the bone marrow contains between 10% and 19% blast cells and the number also increases (to between 5% and 19%) in the circulating blood. The number of red cells, white cells and platelets in the circulating blood is often reduced and there is a greater likelihood of transforming to acute myeloid leukaemia.

Myelodysplastic/myeloproliferative neoplasms (MDS/MPN)

These are a group of diseases that have characteristics of both myelodysplastic (abnormal bone marrow cells producing too few blood cells) and myeloproliferative (abnormal bone marrow cells producing too many blood cells) diseases. These include chronic myelomonocytic leukaemia (CMML), juvenile myelomonocytic leukaemia (JMML), atypical chronic myeloid leukaemia (aCML) and myelodysplastic/myeloproliferative diseases unclassifiable (MDS/MPD-U).

Chronic myelomonocytic leukaemia (CMML) is an example of MDS with higher than normal white counts in the blood, mainly due to abnormal monocytes and myelocytes (immature white cells). Chemotherapy drugs given orally, or sometimes by injection, may be used to control the level of the white counts.

PROGNOSIS

A prognosis is an estimate of the likely course of a disease and the chances of curing or controlling it for a given time.

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

If you have MDS your overall prognosis depends on many factors. A scoring system known as the International Prognostic Scoring System (IPSS) has been developed to provide an estimate of how your disease might progress. There are many other scoring systems that are used in some people with MDS such as the Revised International Prognostic Scoring system (R-IPSS). However the IPSS is the scoring system currently used in Australia.

The International Prognostic Scoring System (IPSS)

The severity of your disease is determined using the International Prognostic Scoring System (IPSS). This system is used to help predict the likely course of your disease, and the risk of your disease transforming to acute myeloid leukaemia.

Using this system, different factors including your blood cell count at diagnosis, the percentage of blast cells seen in your bone marrow and the types of chromosomal abnormalities detected are given individual scores, which are then tallied to give your overall score.

*Depending on your score, you will be regarded as being in one of the following four risk categories: **low, intermediate-1, intermediate-2 or high risk**. Those in the low risk category are less likely to transform to leukaemia and they are expected to survive longer, with some people living to their normal life expectancy. Those in higher risk categories are at greater risk of developing leukaemia and are generally expected to have a reduced survival time.*

For some people MDS remains stable for many years causing few symptoms. Unfortunately for others, it can progress rapidly, transforming into leukaemia. Signs that the disease is progressing include more frequent infections, spontaneous skin bruises and other bleeds (usually gums and nose) and the need for more frequent blood transfusions.

TREATMENT FOR MDS

The treatment chosen for your disease depends on several factors; the exact type of MDS you have, your age, other prognostic factors, and your general health.

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

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

Standard therapy

Standard therapy refers to types of treatment which are commonly used and have been tried and tested both in clinical trials and through long-term clinical use and have been proven to be reasonably safe and effective.

Regular observation (Watch and Wait)

Many people, particularly in the early stage of disease remain very well, living a relatively normal life for a long time without any treatment. At this stage the bone marrow is still relatively healthy. If you are at this stage, your doctor may simply recommend regular checkups to carefully monitor your health.

There are several treatment options available for you if you develop symptoms from your disease.

Supportive care

Supportive care is the mainstay of treatment for the majority of people with MDS. This involves making every effort to improve your quality of life by relieving any symptoms you might have and by preventing and treating any complications that arise from your disease or treatment.

Blood transfusions, antibiotics and, in some cases, the use of growth factors, which promote the production of blood cells in your bone marrow, are all important elements of supportive care.

Blood transfusions

If symptoms of anaemia are interfering with your normal daily activities, your doctor may recommend that you have a blood transfusion. Platelet transfusions are sometimes given to prevent or treat bleeding (for example a persistent nose bleed).

You do not need to stay overnight in a hospital for a blood or platelet transfusion, they are usually given in the clinic or outpatient department of the hospital. Transfusions are relatively safe and they don't usually cause any serious complications. Nevertheless you will be carefully monitored throughout the transfusion. In the meantime, remember to alert the nurse if you are feeling hot, cold, and shivery or in any way unwell, as this might indicate that you are having a reaction to the transfusion. Steps can be taken to minimise these effects and ensure that they don't happen again.

Side-effects of repeated transfusions

You can ask your doctor or nurse for a more detailed booklet outlining the side effects of blood transfusion.

Risk of infection

All blood donors and each unit of blood are screened separately to ensure that harmful viruses are not passed on in a transfusion.

Transfusion reactions

Careful checks are made both at the blood bank and beside you to ensure that the transfusion you are receiving is compatible with your blood type. However, people can become sensitised to red cell and platelet transfusions over time and this can cause, in some cases, a minor transfusion reaction such as a fever or rash. These reactions are usually caused by a small number of white cells present in bags of donated blood and platelets. More recently, these reactions have been dramatically reduced by the use of filtered blood.

Fluid overload

Each bag of blood adds between 250- 400 mls of fluid to your circulation which puts an extra load on the heart. The body usually adjusts to this by producing more urine. Elderly people's heart and kidneys may have difficulty in coping with this relatively sudden increase which can make you feel a bit breathless. To prevent or to treat this, a drug is often given to help you pass urine. The nurse

will ensure that you are told when this is given and that you have easy access to toilet facilities as the drug can be very effective and may start working within 15 minutes.

Iron overload

Over time, repeated blood transfusions can lead to a build-up of high levels of iron in key parts of the body, especially the heart, liver and pancreas. This is called iron overload. Unfortunately, the body has no way to get rid of any extra iron. Your doctor will be able to tell you if this build-up of iron is happening based on the number of transfusions that you may have had and a simple blood test (serum ferritin test). You may want to ask your doctor about iron overload precautions after you have had more than ten bags of blood transfused. If needed, a special drug can be given which binds the iron and helps to safely remove it from the body.

Antibiotics

When your white cell count is low you are at risk of developing an infection. If you develop an infection it is important that you are treated promptly with antibiotics. Don't hesitate to contact your doctor or hospital if you develop any of the following:

- a temperature of 38° C and/or an episode of shivering, called a rigor (where you shake uncontrollably)
- coughing or shortness of breath
- a sore throat and/or a head cold
- passing urine frequently or pain when passing urine
- if you are feeling generally unwell
- if you cut, or otherwise injure yourself
- if you are bleeding (for example blood in your urine, stools, sputum, bleeding gums or a persistent nose bleed) or bruising easily

Please also advise your doctor if any surgery or dental work is planned by another medical practitioner as advice may be required from your haematologist as to the best supportive treatment with red cells, platelets and antibiotics to ensure that your surgery is completed successfully without problems due to your MDS.

Growth factors

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

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

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

Standard drug therapies

Azacitidine

Azacitidine may be given for people with intermediate II or high grade MDS. At the doses used to treat MDS, azacitidine works in a different way to standard chemotherapy and has few of the side-effects of standard chemotherapy. It works by restoring a more normal pattern of gene activity which allows the bone marrow to function more effectively. It is given by injection under the skin, usually for seven days every four weeks. Side effects include a temporary reduction in the blood counts and reactions at the sites of injection.

Because azacitidine begins to become less active soon after it is made up, injections usually can't occur at home and it needs to be given at a hospital day clinic. In patients with higher-risk MDS, azacitidine has been shown to improve survival, increase blood counts, reduce the requirement for blood transfusions and reduce symptoms of the disease. A demonstrated improvement in your MDS can take six months or more to occur with this medication.

Chemotherapy

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

In general, chemotherapy is only used in MDS in situations when there is a need to control a rising white cell count or if the MDS is transforming or has transformed into leukaemia. Chemotherapy is also sometimes given to treat a subtype of MDS called chronic myelomonocytic leukaemia (CMML), which is characterised by a higher than normal white cell count in the blood.

The aim of chemotherapy is to reduce the number of blast cells in your bone marrow and by doing so, allowing the remaining normal stem cells to make normal red blood cells, white blood cells and platelets.

Chemotherapy may be administered in three different situations in MDS.

1. Low-dose oral chemotherapy or cytotoxic therapy for CMML

Low doses of oral chemotherapy (tablets) can be very effective at controlling a high white cell count. Hydroxyurea is an example of an oral chemotherapy drug used in the treatment of a CMML. Hydroxyurea can be taken in capsule form. It is usually very well tolerated and does not usually cause nausea (feeling sick) or significant hair loss, although it can cause dry skin.

The dose of the chemotherapy drug can be adjusted to the response of the white cells and also the response of other blood cells such as red cells and platelets. For example, sometimes a balance has to be made between the effect on lowering the white cell count and the increase in anaemia and thrombocytopenia caused by the drug. Blood counts are monitored frequently while you are receiving chemotherapy.

2. Low-dose intravenous chemotherapy or cytotoxic therapy for high-risk MDS/AML

Low-dose intravenous chemotherapy can be used to control a rising blast count in your blood. This is often seen when MDS is transforming to acute leukaemia. In this case chemotherapy is often given in combination with regular blood and platelet transfusions.

The aim of this treatment is to control the leukaemia while avoiding any severe side-effects from chemotherapy. It is hoped that this will enable you to have a reasonable quality of life and continue to live at home, although visits to the chemotherapy day centre or clinic may be necessary two or three times a week.

Some people who receive low dose chemotherapy do as well as others who receive more aggressive therapy. Unfortunately neither approach will produce a cure of the underlying disease. The choice of which course to take will depend on many factors, including your wishes, the nature of your individual disease and your haematologist's advice.

3. Standard-dose chemotherapy for high-risk MDS/AML

People who have MDS that is transforming, or has transformed, into acute myeloid leukaemia*, may benefit from anti-leukaemia therapy if they are fit enough. Not everyone is suitable for this form of treatment, especially if they are elderly or frail. However, older people do respond similarly to younger patients if they are well enough to tolerate the treatment. Unfortunately, even if a complete remission is achieved, most patients will relapse and the leukaemia will reappear, usually within a year. The decision to have this type of treatment needs to be discussed by you and your family in detail with your doctor.

This treatment is given in hospital and the side-effects can be more severe. If you are having chemotherapy your doctor and nurse will tell you about the side-effects you might experience and how they can be best managed.

**There is a separate Leukaemia Foundation booklet called 'Understanding Acute Myeloid Leukaemia' which provides more details of this type of chemotherapy.*

Potential side effects of chemotherapy

feeling sick - nausea and vomiting

feeling tired and weak

hair loss (alopecia) and thinning

mouth problems

diarrhoea or constipation

skin problems

drop in blood counts

fertility problems

increased sensitivity to the sun

Look good...Feel Better is a free community service program that helps to improve the wellbeing and confidence of people undergoing treatment for any sort of cancer. The program is available free for women, men and teens to help manage the appearance-related side-effects caused by cancer treatment. For more information visit www.lgfb.org.au or 1800 650 960.



Lenalidomide

Lenalidomide is taken as a capsule that needs to be taken on a daily basis. It has been shown to decrease the requirement for blood transfusions, particularly in patients with myelodysplastic syndrome with the isolated del(5q) chromosome subtype. In Australia, lenalidomide is subsidised for the treatment of lower risk MDS where there is a del(5q) and regular blood transfusions may be required.

Stem cell transplantation*

Stem cell transplantation (also called a bone marrow transplant) using a suitably matched donor, is the only potential cure for MDS. This treatment carries significant risks and is only suitable for a small minority of younger patients with MDS.

A standard stem cell transplant involves giving very high doses of chemotherapy, sometimes in combination with radiotherapy, in an attempt to completely destroy the abnormal stem cells in your bone marrow. These cells are then replaced with healthy stem cells which have been donated, usually from a brother or sister who has the same tissue type as yours. This is called an allogeneic (donor) stem cell transplant. In some cases the donor is not a family member, but has a similarly matched tissue type. This type of transplant is called a matched unrelated donor transplant (MUD) or volunteer unrelated donor transplant (VUD).

A newer approach in stem cell transplantation involves using less intensive doses of chemotherapy. This approach may be suitable for selected patients older than 50 years of age. More moderate doses of chemotherapy are used to destroy enough abnormal stem cells in the bone marrow and suppress the patient's immune system enough for it to accept the new, donated stem cells. This is called a reduced intensity transplant, mini transplant, or a mini-allogeneic (mini allo) stem cell transplant.

**There are separate Leukaemia Foundation booklets called 'Understanding Autologous Transplants - A guide for patients and families' and 'Understanding Allogeneic Transplants - A guide for patients and families' that provide more detail on these types of treatments.*

New and experimental drug therapies

There are several new approaches being developed for the treatment of MDS. These include new chemotherapy drugs, targeted therapies, biological modifiers and drugs which harness the power of the immune system to help fight disease. Side effects vary according to the type of drug used. Some of these drugs are available in Australia, others are only available on clinical trials.

For younger patients with hypoplastic MDS, immunosuppressive (drugs to reduce your immune system) therapies such as anti thymocyte globulin and cyclosporine may lead to an improvement in blood counts.

In a small number of patients with CMML who have a specific genetic abnormality involving the gene for platelet derived growth factor receptor beta (PDGFRB), a targeted therapy called imatinib may be used.

Your doctor will be able to discuss with you all of the suitable treatment options.

New and experimental drugs for MDS

Angiogenesis Inhibitors / Immune modulators (inhibit growth factors and new blood vessels)

Thalidomide
Lenalidomide
Arsenic Trioxide

Histone Deacetylase Inhibitors (inhibit abnormal gene activity)

Phenylbutyrate
Valproic acid, panobinostat

Farnesyl Transferase Inhibitors (inhibit abnormal cell growth signals)

Tipifarnib
Lonafarnib

Polo-like kinase/Phosphoinositide-3 kinase Inhibitors (inhibit abnormal gene activity)

Rigosertib

Clinical trials

These trials (also called research studies) test new treatments and compare them to established treatments both to see if the new treatment works better and to compare the side effects. Some clinical trials are randomised, so that some patients will be receiving the new treatment and others the old treatment. Clinical trials are important because they provide vital information about how treatment could be improved. Clinical trials sometimes give access to new therapies that have been trialed elsewhere but are not yet available locally.

If you are considering taking part in a clinical trial make sure that you understand the reasons for the trial and what it involves for you. You also need to understand the benefits and risks of the trial before you can give your consent. Talk to your doctor who can guide you in making the best decision for you.

Informed consent

Giving an **informed** consent means that the potential risks and potential benefits of a proposed procedure or treatment have been fully explained to you. You will then be asked if you are willing to comply with the requirements of the trial, including treatments, tests and the collection of relevant information about you and your disease. If you feel happy that you have adequate information to make such a decision, you may then give your informed consent.

You should never feel compelled to be in a clinical trial. The decision to participate must always be voluntary, after you have received all the necessary information. Even after you have agreed to be in a clinical trial and signed the consent form, you can change your mind, whether or not trial treatment has already started.

“How can I help with blood cancer research?”

The Australasian Leukaemia and Lymphoma Group (clinical trials research group) has established a national Leukaemia and Lymphoma Tissue Bank at the Princess Alexandra Hospital in Brisbane. The Tissue Bank is a temperature controlled facility for storing clinical tissue samples to be used in approved research into leukaemia, lymphoma, myeloma and related blood disorders. Current research focuses on understanding the development of cancers, why different patients respond differently to current treatments and more effective therapies, especially those being assessed in clinical trials. The clinical tissue samples used for this research come from blood and bone marrow samples from patients' routine testing and from samples taken for monitoring during clinical trials.

In order to donate your blood and/or bone marrow samples to the Tissue Bank you will need to sign a **consent form** at the time of your diagnosis. This can be obtained from your clinician. Be assured, donating does not involve any additional procedures, it simply involves saving and storing in the Tissue Bank any excess blood or bone marrow extracted during your routine tests. Samples are also welcomed from relapsed patients at re-diagnosis.

The donation of your tissue sample is an invaluable way to support blood cancer research and could bring us closer to finding a cure. Tissues from blood cancer patients are precious materials for researchers because these cancers are relatively rare and are vital for finding cures.

Fertility

The following information has been included for people who are concerned about the effects of chemotherapy and radiotherapy on their ability to have children in the future.

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

In women, some types of chemotherapy and radiotherapy can cause varying degrees of damage to the normal functioning of the ovaries. In some cases this leads to menopause (change of life) earlier than expected. In men, sperm production can be impaired for a while but the production of new sperm may return to normal in the future. There are some options for preserving your fertility, if necessary, while you are having treatment for MDS. These are described below.

Protecting your fertility - Men

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

If possible, semen should be donated on more than one occasion.

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

Protecting your fertility - Women

Protecting your fertility - Women

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

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

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

To date, egg storage and ovarian tissue storage are techniques which remain under investigation.

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

It is important to understand that the methods are still experimental and for many reasons achieving a pregnancy and subsequently a baby is not guaranteed by using any of them. In addition, some are time consuming and costly while others may simply not be acceptable to you or your partner. For aggressive disease, the best treatment option may require immediate chemotherapy and fertility options may be limited.

Early menopause

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

Hormone changes can lead to many of the classic symptoms of menopause including menstrual changes, hot flushes, sweating, dry skin, vaginal dryness and itchiness, headache and other aches and pains. Some women experience decreased sexual drive, anxiety and even depressive symptoms during this time. It is important that you discuss any changes to your periods with your doctor or nurse. He or she may be able to advise you, or refer you on to a specialist doctor (a gynaecologist) or clinic that can suggest appropriate steps to take to reduce your symptoms.

Body image, sexuality and sexual activity

It is likely that the diagnosis and treatment of MDS will have some impact on how you feel about yourself as a man or a woman and as a 'sexual being'. Hair loss, skin changes, and fatigue can all interfere with feeling attractive. As we mentioned previously, Look Good... Feel Better is a free community service that runs programs on how to manage the appearance-related side-effects of cancer treatments. You might like to visit their website at

www.lgfb.org.au or freecall them on 1800 650 960.

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

It is usually recommended that you or your partner do not become pregnant as some of the treatments given might harm the developing baby. As such you need to ensure that you or your partner use a suitable form of contraception. Condoms (with a spermicidal gel) offer good contraceptive protection as well as protection against infection or irritation. Partners are sometimes afraid that sex might in some way harm the patient. This is not likely as long as the partner is free from any infections and the sex is relatively gentle. Finally, if you are experiencing vaginal dryness, a lubricant can be helpful. This will help prevent irritation. If you have any questions or concerns regarding sexual activity and contraception don't hesitate to discuss these with your doctor or nurse, or ask for a referral to a doctor or health professional who specialises in sexual issues.

Complementary therapies

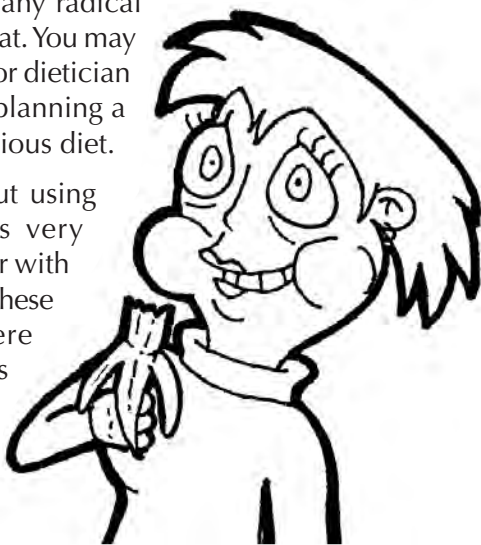
Complementary therapies are not considered standard medical therapies. Many people however find that they are helpful in coping with their treatment and recovery from disease. There are many different types of complementary therapies. These include yoga, exercise, meditation, prayer, acupuncture, relaxation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

Complementary therapies should 'complement' or assist with recommended medical treatment. **They should not be used as an alternative to medical treatment for MDS.** It is important to realise that no complementary or alternative treatment alone has proven to be effective against MDS. It is also important to let your doctor or nurse know if you are using any complementary or alternative treatments, in case they interfere with effectiveness of chemotherapy or other treatments you may be having.

Nutrition*

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

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



**The Leukaemia Foundation has a booklet called 'Eating Well - A Practical Guide for People living with Leukaemias, Lymphomas, Myeloma and Related Blood Disorders' that provides further detail.*

MAKING TREATMENT DECISIONS

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

Before going to see your 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 treating doctor (haematologist) will spend time discussing with you and your family what he or she feels is the best option for you. Feel free to ask as many questions as you need to, at any stage. You are involved in making important decisions regarding your well-being. You should feel that you have enough information to do this and that the decisions made are in your best interests. Remember, you can always request a second opinion if you feel this is necessary.

INFORMATION AND SUPPORT*

People cope with a diagnosis of MDS in different ways and there is no right or wrong or standard reaction. For some people the diagnosis can trigger any number of emotional responses ranging from denial to devastation. It is not uncommon to feel angry, helpless and confused. Naturally people fear for their own lives or that of a loved one. On the other hand, people who do not currently require treatment may wonder if they are sick at all.

It is worth remembering that information can often help to take away the fear of the unknown. It is best for patients and families to speak directly to their doctor regarding any questions they might have about their disease or treatment. It can also be helpful to talk to other health professionals including social workers or nurses who have been specially educated to take care of people with diseases like MDS. Some people find it useful to talk with other patients and family members who understand the complexity of feelings and the kinds of issues that come up for people living with an illness of this nature.

There may be a MDS support group in your state or territory. You may wish to contact the Leukaemia Foundation for more information.

If you have a psychological or psychiatric condition please inform your doctor and request additional support from a mental health professional, especially if your treatment includes the use of steroid therapies.

Many people are concerned about the social and financial impact of the diagnosis and treatment on their families. Normal family routines are often disrupted and other members of the family may suddenly have to fulfil roles they are not familiar with, for example cooking, cleaning, doing the banking and taking care of children.

There are a variety of programs designed to help ease the emotional and financial strain created by blood cancers and related disorders. The Leukaemia Foundation is there to provide you and your family with information and support. The Leukaemia Foundation has a number of Support Services Coordinators who can assist you in many ways. Contact details for your state office are provided on the back of this booklet.

**There is a separate Leukaemia Foundation booklet called 'Living with Leukaemias, Lymphomas, Myeloma and Related Disorders'. This booklet addresses the impact of the diagnosis, family matters, support, survivorship, and other general issues around treatment.*

USEFUL INTERNET ADDRESSES

- Leukaemia Foundation
www.leukaemia.org.au
- American Cancer Society
www.cancer.org
- Aplastic Anaemia & MDS International Foundation, Inc.
www.myelodysplasticsyndromes.org or www.aamds.org
- Cancer Council of Australia
www.cancer.org.au
- Clinical Trials
www.australiancancertrials.gov.au
- Leukaemia Foundation's - Online Support Group
www.talkbloodcancer.com
- Leukemia & Lymphoma Society of America
www.leukemia-lymphoma.org
- Leukaemia Research Fund (UK)
www.lrf.org.uk
- Look Good ... Feel Better program
www.lgfb.org.au
- Macmillan Cancer support
www.macmillan.org.uk
- MDS UK Patient Support Group
<http://www.mdspatientsupport.org.uk/>
- MDS Foundation
www.mds-foundation.org.
- The MDS Beacon
www.mdsbeacon.com

GLOSSARY OF TERMS

Alopecia

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

Anaemia

A reduction in the number of red blood cells or 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.

Bone marrow

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

Blast cells

Immature blood cells normally found in the bone marrow in small numbers.

Blood count

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

Cancer

A malignant disease characterised by uncontrolled growth, division, and accumulation of abnormal cells. There is invasion into other tissues 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.

Central Venous Catheter (CVC)

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

Chemotherapy

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

Chromosomes

Chromosomes are made up of coils of DNA (deoxyribonucleic acid). DNA carries all the genetic information for the body in sequences known as genes. There are approximately 20,000 different genes carried on 46 chromosomes. The chromosomes are contained within the nucleus of a cell.

Cytogenetic tests (studies)

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

Growth factors and cytokines

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

Haemopoiesis

The processes involved in blood cell formation.

Haematologist

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

Immune system

The body's defence system against infection and disease.

Leukaemia

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

Lymphocyte

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

Mucositis

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

Neutropenia

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

Neutrophils

Neutrophils are the most common type of white blood cell. They are needed to effectively fight infection, especially bacterial and fungal infections.

Prognosis

An estimate of the likely course of a disease.

Radiotherapy (radiation therapy)

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

Relapse

The return of the original disease.

Remission (or Complete Remission)

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

Stem cells

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

Syndrome

A characteristic collection of medical symptoms and signs; for example, the syndrome in myelodysplastic syndrome, refers to the fatigue due to anaemia, increased tendency to infections and increased bruising which are all features of myelodysplasia.

White blood cells

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

Please send me a copy of the following information booklets and newsletters:

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

Or information about:

- The Leukaemia Foundation's Support Services
- Regular salary deduction scheme
- National Fundraising Campaigns
- Volunteering
- Bequests (including the Leukaemia Foundation in your Will)

Name:

Street or Postal Address:

Suburb:

State/Postcode

Email: Tel: (.....)

Please send to:

Leukaemia Foundation, GPO Box 9954, In Your Capital City

or Freecall 1800 620 420

or email: info@leukaemia.org.au

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







Leukaemia Foundation

VISION TO CURE
MISSION TO CARE

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

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

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

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

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

January 2014

Printed by:



This paper is:

