

# Myelodysplastic Syndromes (MDS)

A guide for patients, families & whānau



our mission is to care, our vision is to cure

## Contents

Introduction	2
Leukaemia & Blood Cancer New Zealand	3
Bone marrow, stem cells and blood cell formation	5
What is MDS?	8
How common is MDS and who gets it?	9
What causes MDS?	9
What are the symptoms of MDS?	10
How is MDS diagnosed?	11
Types of MDS	13
Prognosis	15
How is treatment decided?	16
Making treatment decisions	17
Treatment for MDS	18
Supportive care	21
Reproductive health	27
Body image, sexuality & sexual activity	29
Social and emotional effects	30
Useful internet addresses	31
Glossary of terms	32

## Introduction

This booklet has been written to help you and your family or whānau understand more about myelodysplastic syndromes (MDS).

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

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

We have used some medical words and terms that you may not be familiar with. Their meaning is either explained in the text, in the glossary of terms at the back of this booklet, or in the 'Dictionary of Terms' booklet available from Leukaemia & Blood Cancer New Zealand.

Some people may require more information than is contained in this booklet. We have included some internet addresses that you might find useful. In addition, many of you will receive written information from the doctors and nurses at your treatment centre.

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

We hope that you find this booklet useful. There is a feedback form at the back of this booklet, please feel free to fill this in and return it to us to assist in the production of future editions.

There is a separate information booklet called 'Myeloproliferative Disorders – a guide for patients, families & whānau' available from Leukaemia & Blood Cancer New Zealand.

## Acknowledgements

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

Leukaemia & Blood Cancer New Zealand also gratefully acknowledges Dr Robert Weinkove (Wellington Hospital) and Dr Hilary Blacklock (Middlemore Hospital) for their assistance with the development of this booklet

## Leukaemia & Blood Cancer New Zealand

Leukaemia & Blood Cancer New Zealand (LBC) is the only organisation in New Zealand dedicated to supporting patients and their families living with leukaemia, lymphoma, myeloma and related blood conditions.

Since 1977, our work has been made possible through our fundraising events and the generous support we receive from individuals, companies, trusts and grants. We do not receive government funding.

LBC manages the New Zealand Bone Marrow Donor Registry (NZBMDR), which works towards finding matched volunteer donors from New Zealand or overseas for New Zealand patients who need a bone marrow or stem cell transplant and who do not have a family donor. The registry maintains information on New Zealand donors and has access to a worldwide database of over 20 million donors.

## Patient Support

Leukaemia & Blood Cancer New Zealand's Support Services provide personalised support programmes for patients and their families. This can include regular visits, phone or email contact, as well as face to face education and support programmes, and an online information forum. We also provide a toll free number for advice and support.

## Research

Research plays a critical role in building a greater understanding of blood cancers and conditions. LBC supports and funds investigation into these conditions. Improved treatments for patients can lead to less side effects and increased survival rates.

## Information

We provide vital information to patients, families, health professionals and the community to improve understanding about blood cancers and conditions.



## Awareness

We work to increase public knowledge of blood cancers and conditions. This is achieved through specifically focused campaigns for the public, health professionals and health agencies.

## Advocacy

We represent the needs of patients and their families to the government, related agencies and other relevant organisations.

## Contacting us

Leukaemia & Blood Cancer New Zealand provides services and support throughout New Zealand. Each person's experience of living with a blood cancer or condition is different. Living with leukaemia, lymphoma, myeloma or a related blood condition is not easy, but you do not have to do it alone.



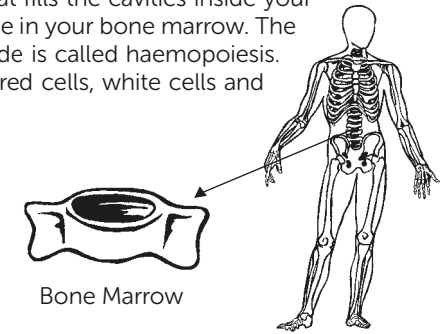
Call **0800 15 10 15** to speak to a local Support Services Coordinator or to find out more about the services offered by Leukaemia & Blood Cancer New Zealand. Alternatively, contact us via email by sending a message to [info@leukaemia.org.nz](mailto:info@leukaemia.org.nz) or by visiting [www.leukaemia.org.nz](http://www.leukaemia.org.nz).

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

## Bone marrow, stem cells & blood cell formation

### Bone marrow

Bone marrow is the spongy tissue that fills the cavities inside your bones. All of your blood cells are made in your bone marrow. The process by which blood cells are made is called haemopoiesis. There are three types of blood cells; red cells, white cells and platelets.

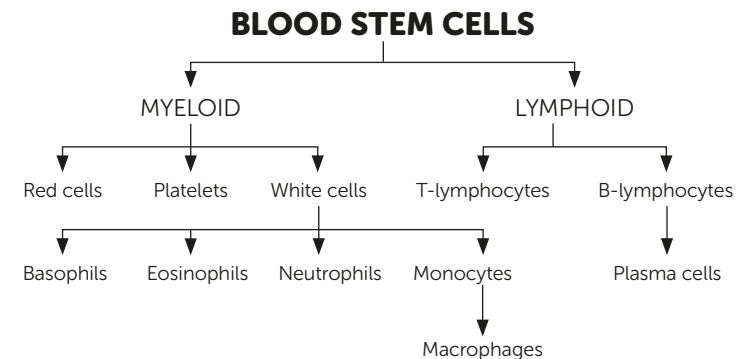


As an infant, haemopoiesis takes place in the centre of all bones. As an adult, fewer new blood cells are needed - the marrow space in the arms and legs is replaced by fat, and active bone marrow is limited to the hips, pelvis, ribs, breastbone (sternum) and back bones (spine or vertebrae).

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

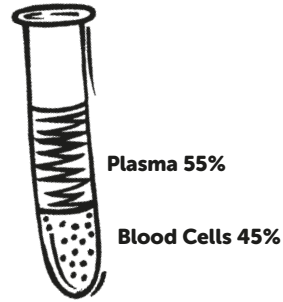
**Myeloid** ('my-loid') stem cells develop into red cells, white cells (neutrophils, eosinophils, basophils and monocytes) and platelets.

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



## Blood

Blood consists of blood cells and plasma. Plasma is the straw-coloured fluid part of the blood which contains important proteins, such as clotting factors. Plasma carries these essential nutrients, including blood cells, around the body.



## Blood cells

### Red cells and haemoglobin

Red cells contain haemoglobin (Hb) which transports oxygen from the lungs to all parts of the body. Haemoglobin also carries carbon dioxide to the lungs where it can be breathed out. Haemoglobin is what makes red blood cells red.

The normal haemoglobin range for a man is between 130 - 170 g/L

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

Red cells are by far the most numerous blood cells and the proportion of the blood that is occupied by these is called the haematocrit. A low haematocrit suggests that the number of red cells in the blood is lower than normal.

The normal haematocrit range for a man is between 0.40 – 0.52  
(40 – 52%)

The normal haematocrit range for a woman is between 0.36 – 0.46  
(36 – 46%)

Anaemia is a condition caused by a reduction in the haemoglobin level of the red blood cells. The severity of the anaemia relates to the level of measured haemoglobin with the lower the level of haemoglobin, the greater the severity 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 blood cell transfusion may be given to increase the red cell numbers and therefore the haemoglobin level.

## White cells

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

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

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

The normal adult white cell range is between  $4.0 - 11.0 \times 10^9/L$

Neutropenia is the term given to describe a lower than normal neutrophil count. If you have a neutrophil count of less than 1.0 ( $1.0 \times 10^9/L$ ) you are considered to be neutropenic and at risk of developing frequent and sometimes severe infections.

The normal adult neutrophil range is between  $2.0 - 7.5 \times 10^9/L$

## Platelets

Platelets are disc-shaped cellular fragments that circulate in the blood and play an important role in clot formation. They help to prevent bleeding. If part of your body is injured or cut or a blood vessel is damaged, the platelets gather at the site of the injury, stick together and form a plug to help stop the bleeding.

The normal adult platelet range is between  $150 - 400 \times 10^9/L$

Thrombocytopenia is the term used to describe a low platelet count. If your platelet count is low, you are at higher risk of bleeding and will tend to bruise more easily. Platelet transfusions are sometimes given to bring the platelet count back to a higher level. In certain situations, especially when patients are receiving treatment, platelets may be transfused if the blood level falls below  $10 \times 10^9/L$ .

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

## Growth factors and cytokines

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

Many growth factors can be made in the laboratory (synthesised) and are available for use in people with blood disorders. For example, granulocyte-colony stimulating factor (G-CSF) stimulates the production of white cells called neutrophils, while erythropoietin (EPO) stimulates the production of red cells and thrombopoietin (TPO) stimulates the production of platelets. See page 24 for more about growth factors.

## What is MDS?

Myelodysplastic syndromes (MDS) are a group of diseases which all affect, to a greater or lesser extent, the production of normal blood cells in the bone marrow. MDS is also sometimes referred to as myelodysplasia.

In MDS, abnormal bone marrow stem cells produce abnormal blood cells which do not grow properly. The blood cells that do grow may be abnormal in 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 results in lower numbers of normal functioning red blood cells, white blood cells and platelets being produced. This means that people with MDS often have a very active bone marrow but a low number of circulating blood cells. Without enough red blood cells, white blood cells and platelets you can become fatigued, more susceptible to infections, and can 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 MDS.

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 have only anaemia, or they might have low numbers of white cells and/or platelets. In many cases they have few, if any, troubling symptoms from their disorder. In more severe cases, the lack of circulating blood cells is more pronounced, causing more symptoms.

In some cases of MDS, the number of abnormal bone marrow stem cells (called blasts) is increased. Patients with an increased number of blasts in the bone marrow are at risk of progression to acute myeloid leukaemia (AML).

## How common is MDS and who gets it?

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, or it may be picked up incidentally during a routine blood test. MDS is not contagious; you cannot 'catch' MDS by being in contact with someone who has the disease and it is not inherited or passed on within families.

Over 85 per cent of cases occur in people over the age of 60 years with males more affected than females, however MDS can occur at any age, including very occasionally in children.

## What causes MDS?

Defects arise in the bone marrow stem cells which result in MDS. Why these happen in a particular person at a particular time is difficult to understand, although the effects of ageing on cell growth appear to play a major role. There are also some recognised factors which may put some people at a higher risk of developing MDS. These are called risk factors or predisposing factors and they are described below.

MDS occurs as a result of mutations (or changes) in one or more of the genes that control blood cell development. This change results in the abnormal growth or survival of blood stem cells. The mutations are preserved when the affected stem cell divides, producing a 'clone'; that is a group of identical cells all with the same defect. This is why MDS can be described as a clonal blood stem cell disorder.

Mutations in dividing cells occur all the time and cells have clever ways of correcting these mutations. However the longer we live, the more chance we have of acquiring mutations that manage to escape these safeguards. 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. Any process which damages genes and leads to mutations may have a role in the development of MDS. Some known risk factors include:

**Ageing:** As mentioned above, 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 the many chemicals in tobacco smoke (which include benzene produced by burning the tobacco) may increase the risk of developing MDS.

**Chemotherapy:** People previously treated for cancer or other conditions with certain cytotoxic chemotherapy drugs 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 often associated with different mutations than those that occur in spontaneous MDS, and has a less favourable prognosis. The time between exposure to the drugs and development of MDS may be over 10 years.

**Radiation:** Previous radiotherapy, or accidental exposure to high levels of environmental gamma radiation, is associated with an increased risk of MDS, which in some cases may not be apparent for up to 40 years.

**Rare hereditary or congenital causes:** People with certain congenital disorders such as Bloom syndrome, Down 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 have no symptoms at all and their MDS is picked up incidentally during a routine blood test. In other cases people go to see their general practitioner (GP) because of troubling symptoms. 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:

1. Lack of red cells, or anaemia. This can lead to:
  - Persistent tiredness and fatigue
  - Weakness
  - Shortness of breath with minimal exercise
  - Looking pale
  - Dizziness
  - Angina in patients with heart disease
2. Abnormal white cell function, usually with low white cell counts. This can lead to:
  - Recurring infections, especially chest infections
  - Fevers
  - Sore mouth due to mouth ulcers
3. Abnormal platelet function, often with low platelet counts. This can lead to:
  - Easy bruising (purpura)
  - Petechiae – a rash of small red dots, often seen on the lower limbs often seen initially due to bleeds from small blood vessels (capillaries) within the skin
  - Tendency to bleeding 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.

## Which doctor?

If your GP suspects that you might have MDS that requires treatment, you will be referred on to another specialist doctor called a haematologist for further tests and treatment. A haematologist specialises in the care of people with diseases of the blood, bone marrow and immune system.

For people with MDS not needing treatment, it is common to remain under the care of your GP, with regular blood tests as required.



## How is MDS diagnosed?

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

### Full blood count

The first step in diagnosing MDS is 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 a blood film is examined 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 MDS. Other blood tests will be done to rule out other causes of low blood counts, for example; low iron, folate and vitamin B12 levels, certain viral infections, and abnormal kidney function.

### Bone marrow biopsy or examination

A bone marrow biopsy involves taking a sample of bone marrow, usually from the back of the pelvis (hip bone) or rarely from the sternum (breast bone). This is then sent to the laboratory for examination under the microscope, to check if there are any abnormal cells present and to see how well the bone marrow is functioning.

The bone marrow examination may be done in the hospital or outpatient clinic with a local anaesthetic or, in selected cases, under sedation. 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 thin needle is inserted through the skin and the 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 sucked out - this is called a 'bone marrow aspirate'.

Then a slightly wider needle is used to obtain a thin core of bone marrow to provide information about the structure of the bone marrow and bone - this is known as a 'bone marrow trephine'.

If a sedative is used you might feel drowsy afterwards, and you will be asked to take a family member or friend along who can take you home. A small dressing or plaster over the biopsy site can be removed the next day.

There may be some mild bruising, or discomfort which usually is managed effectively by paracetamol. More serious complications such as bleeding or infection are rare.

### Special tests

Special laboratory tests may be undertaken using blood or bone marrow biopsy samples.

#### Cytogenetic ('cy-to-gen-etic') tests

Chromosomes are the organised structures of DNA inside our cells. A single piece of DNA contains many genes, which are our body's blueprint for life. Cells which have certain types of changes or mutations in their genetic make-up can be detected by cytogenetic testing. The two main types of tests are chromosome analysis, which examines chromosomes under a microscope, and FISH (florescent in-situ hybridisation), which 'paints' the commonly abnormal genes with fluorescent dyes.

Certain cytogenetic mutations, such as missing, extra or abnormal chromosomes help to confirm the sub-type of myelodysplastic syndrome you have, the likely prognosis of your disease and the best way to treat it. Note: these mutations develop as your cells age, so this type of chromosomal change is only found in the cancerous cells and is not inherited (i.e. it is not passed down from parent to child).

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

Immunophenotyping, or flow cytometry, is a laboratory technique used to distinguish MDS from other similar diseases, such as leukaemia. This technology uses fluorescent probes to label markers, called antigens, found on the surface of cells. Specific combinations of antigens may be found on the abnormal cells of MDS, and this can help to confirm the diagnosis.

Antigens are commonly referred to as 'clusters of differentiation' or CD antigens followed by a number (e.g. CD34, CD16).

### Other tests

Once a diagnosis of myelodysplastic syndrome is made, other tests may be used to assess your general health, particularly if treatment is being considered. They may include:

- chest x-ray
- electrocardiogram (ECG)
- additional blood tests such as liver and renal function tests, serum iron studies
- An echocardiogram (cardiac ultrasound), or a nuclear cardiac scan



Waiting for results can be a very distressing time for you and your family. It may help to talk about your feelings with someone you are close to or feel comfortable with.

### Types of MDS

The current (2008) World Health Organisation's (WHO) classification system recognises several major subtypes of MDS (see below). 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 may help your doctor to recommend the best course of treatment for you.

#### Major subtypes of MDS (based on the WHO classification)

**Refractory cytopenias with unilineage dysplasia (RCUD)** - For example: refractory anaemia (RA). In this type of MDS, one blood cell type (red blood cells, neutrophils or platelets) is most affected, causing anaemia or neutropenia or thrombocytopenia. The bone marrow contains fewer than 5% abnormal bone marrow stem cell (blast) cells and there are none found in the circulating blood. This type of MDS is less likely to transform to leukaemia and treatment is regular observation, growth factor support or blood transfusion only.

**Myelodysplastic syndrome associated with isolated del (5q) chromosome** - Red blood cells are affected, causing anaemia. There are usually fewer than 5% blast cells in the bone marrow and none in the circulating blood. The developing blood cells in the bone marrow display the unique chromosome abnormality del (5q). This is an example of a chromosome abnormality associated with a good prognosis.



**Refractory anaemia with ringed sideroblasts (RARS)** - Similar to refractory cytopenia with unilineage dysplasia (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 are deposited in a way that forms a ring around the nucleus of a developing red blood cell. These are called 'ringed sideroblasts', and can be seen under the microscope.

**Refractory cytopenia with multilineage dysplasia (RCMD)** - Two or more blood cell types are usually affected here, but again the bone marrow contains fewer than 5% blast cells and there are usually none found in the circulating blood.

**Refractory anaemia with excess blasts 1 (RAEB-1)** - One or more blood cell types are affected. The bone marrow contains between 5% and 9% blast cells and there are only a small number of blast cells (fewer than 5%) found in the circulating blood.

**Refractory anaemia with excess blasts 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 there may be between 5% and 19% blasts in the circulating blood. The number of red cells, white cells and platelets in the circulating blood is reduced and there is a greater likelihood of transforming to acute myeloid leukaemia.

**Myelodysplastic/myeloproliferative neoplasms/disorders (MDS/MPD)** - These are a group of diseases that have characteristics of both myelodysplastic syndromes (abnormal bone marrow cells producing too few blood cells) and myeloproliferative disorders (abnormal bone marrow cells producing too many blood cells). 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 cell counts in the blood, mainly abnormal monocytes. If treatment is necessary, chemotherapy drugs may be given orally, or sometimes by injection, to control the level of the white cell count.

**Myelodysplasia (unclassifiable)** - Sometimes myelodysplastic syndromes do not fit exactly into any of the above categories.

## Prognosis

A prognosis is an estimate of the likely course of a disease and the chances of controlling it for a given time. Your doctor is the best person to give you a likely prognosis regarding your MDS.

If you have MDS your overall prognosis depends on many factors. Scoring systems such as the International Prognostic Scoring System (IPSS) and WHO classification-based Prognostic Scoring System (WPSS) have been developed to provide an estimate of how your disease might progress.

In 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), regular fevers and sweats and the need for more frequent blood transfusions.

### International Prognostic Scoring System (IPSS)

The prognosis of your MDS can be estimated using the International Prognostic Scoring System (IPSS) or the revised IPSS (IPSS-R). These systems are used to help predict the risk of your disease transforming to acute myeloid leukaemia and your future outlook, once you have been diagnosed with MDS.

Using these systems, different factors including your blood cell counts 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 combined to give an overall risk group.

The IPSS has four risk categories and the IPSS-R has five risk categories. Those in lower risk categories are less likely to transform to leukaemia and they are expected to live longer. Those in higher risk categories are at greater risk of developing leukaemia and are generally expected to have a shorter survival time.

### WHO classification-based Prognostic Scoring System (WPSS)

The WHO classification-based Prognostic Scoring System (WPSS) provides a scoring system that can be used at any time during the course of your MDS, which reflects changes in your condition. This system is based on the WHO classification, karyotype, and frequency of blood product transfusion, as survival time is shorter in patients who require regular blood transfusions versus those who do not.

The WPSS has five risk categories from very low to very high. People change categories over the course of the disease as it progresses. This can aid in making treatment decisions, particularly in patients with lower-risk MDS, whose condition may be stable for many years.

## How is treatment decided?

The type of treatment you will receive depends on a number of factors. These include the type of disease you have, your age, your general health, the condition of your marrow and whether you would benefit from treatment at this time. Your haematologist will discuss with you the best option for your particular situation.

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

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

### Informed consent

Giving your informed consent means that you understand and accept the risks and benefits of a proposed procedure or treatment; thus you know that you have adequate information to make such a decision.

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

If you have any doubts or questions regarding any proposed procedure or treatment, please do not hesitate to talk to your doctor or specialist nurse again.

### Standard therapy

Standard therapy refers to a type of treatment which is commonly used in particular types and stages of disease. It has been tried and tested (in clinical trials) and has proven to be safe and effective in a given situation.

### Clinical trials

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

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

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

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

## Making treatment decisions

Many people feel overwhelmed at the prospect of having treatment. Having to make decisions about proceeding with recommended treatments can be very stressful. Some people do not feel that they have enough information to make such decisions while others may be 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. 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.

### The best option for you

It is important to remember that everyone is different. For some, aggressive treatment is not considered the best way to treat their disease. Other approaches, such as regular monitoring or supportive care alone, may offer some people just as good or an even better quality of life.

The Haematology Patient Diary, available from Leukaemia & Blood Cancer New Zealand, may be useful for recording details of treatment and making notes from clinic appointments.

### Interpreting services

New Zealand's Health and Disability Code states that everyone has the right to have an interpreter present during a medical consultation. Family or friends may assist if you and your doctor do not speak the same language, but you can also ask your doctor to provide a trained interpreter if using a family member is not appropriate.

## Treatment for MDS

The aim of treatment for most people with MDS is to alleviate symptoms of the condition and improve quality of life. Treatment for MDS may involve the use of the following:

- Regular monitoring
- Supportive care
- Chemotherapy
- Stem cell transplantation
- Treatment with drugs not yet available for general use (e.g. clinical trials)

### Regular monitoring

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 relatively healthy, and your doctor may simply recommend regular check-ups and blood testing to monitor your health. This monitoring may be under the care of your GP.

### 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 symptoms you might have and by preventing and treating complications that arise from your disease or treatment.

Blood transfusions, antibiotics and, in some cases, the use of growth factors to promote the production of blood cells in your bone marrow, are all important elements of medical supportive care. See Page 21 for more details.

### Chemotherapy

Chemotherapy literally means therapy with chemicals. Many chemotherapy drugs are also called cytotoxics (cell toxic) because they kill cells; especially those that multiply quickly like the abnormal bone marrow stem cells (blasts) in MDS.

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 already transformed into acute leukaemia. Chemotherapy is also 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 and may be associated with an enlarged spleen.

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. Your blood counts and general health are monitored more frequently while you are receiving chemotherapy.

Chemotherapy may be administered in three different situations in MDS.

### 1. Low-dose oral chemotherapy for CMML

Low doses of oral chemotherapy (chemotherapy that is taken by mouth) 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 cause nausea (feeling sick) or hair loss, although it may 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 count and the increase in anaemia and lower platelet count caused by the drug.

### 2. Low-dose chemotherapy for high-risk MDS/acute myeloid leukaemia (AML)

Low doses of oral, subcutaneous (beneath the skin) or intravenous chemotherapy can be used to control a rising blast count in the peripheral 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 severe side effects from chemotherapy. It is hoped that this will enable you to have a reasonable quality of life and to continue living at home, although visits to the chemotherapy day centre or clinic may be necessary two or three times a week.

Some patients who receive low-dose chemotherapy have good responses to treatment without the toxicity seen with standard chemotherapy. Unfortunately, although it can prolong life, low-dose chemotherapy cannot cure MDS or acute myeloid leukaemia.

### 3. Intensive chemotherapy for high-risk MDS/AML

People who have MDS that is transforming, or has transformed, into acute myeloid leukaemia, may benefit from intensive anti-leukaemia chemotherapy if they are fit enough. Not everyone is suitable for this form of treatment, especially if they are elderly or frail. 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 haematologist.

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 booklet called 'Acute Myeloid Leukaemia – a guide for patients, families and whānau' available from Leukaemia & Blood Cancer New Zealand.

### Potential side effects of chemotherapy

- feeling sick - nausea and vomiting
- feeling tired and weak
- hair loss and thinning
- mouth problems
- diarrhoea or constipation
- skin problems
- drop in blood counts
- fertility problems

### **Stem cell transplant**

A stem cell transplant (also called a bone marrow transplant) using a suitably matched donor, is the only potential cure for MDS. This treatment carries significant risks however and is only suitable for a very small minority (<5%) of younger patients with MDS (usually under 65 years of age).

A stem cell transplant involves giving 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 that 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).

Because most patients with MDS are older, the stem cell transplants performed are often given using less intensive doses of chemotherapy than would be used in young adults. The theory is that moderate doses of chemotherapy will 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 conditioning (RIC) stem cell transplant.

There is a separate booklet called 'Allogeneic Stem Cell Transplants – a guide for patients, families and whānau' available from Leukaemia & Blood Cancer New Zealand that provides more details of this type of treatment.

### **New and experimental drug therapies**

There are several new approaches being developed for the treatment of MDS. These include new chemotherapy drugs, biological modifiers and immunomodulatory drugs which harness the power of the immune system to help fight disease. Side effects vary according to the type of drug used.

Some examples of newer treatments for MDS are listed below. These drugs are not freely available in New Zealand, but may be given as treatment in a clinical trial. Your haematologist will be able to discuss with you all of the available treatment options suitable for you.

### **New and experimental drugs for MDS**

#### **DNA hypomethylating agents (inhibit abnormal gene activity)**

- Azacitidine
- Decitabine

#### **Immunomodulator and phosphatase inhibitor**

- Lenalidomide (particularly for MDS with del 5q-)

#### **Histone deacetylase inhibitors (inhibit abnormal gene activity)**

- Valproic acid

#### **Kinase inhibitors (inhibit abnormal cell growth signals)**

- Sorafenib
- Rigosertib
- AC220, CEP-701 (inhibitors of FLT3)

### **Supportive care**

Supportive care plays an important role in the treatment of people with MDS. This involves making every effort to improve your quality of life, by relieving symptoms and by preventing and treating complications that arise from your disease or treatment.

Blood transfusions, antibiotics, intravenous fluids and similar treatments, are examples of medical supportive care. Non-medical supportive care may involve complementary therapies, nutrition support, exercise, counselling and similar services. Many aspects of supportive care are things that you can do yourself.

Inform your haematologist if any surgery or treatment is planned by another practitioner, as advice may be required from your haematologist as to the best supportive treatment, such as transfusions, blood tests or other monitoring, to ensure that your treatment happens successfully without problems due to your disease, current or previous treatment.

### **Infection precautions**

Infections are common in MDS patients with low numbers of functioning white blood cells or in others after chemotherapy. While infections can occur anywhere in the body; common sites include the upper and lower respiratory infections (chest infections), urinary tract (kidney infections) and skin. While most infections are caused by bacteria and viruses; fungal and opportunistic infections (infections caused by micro-organisms that are normally harmless in healthy people) are also seen. You may be prescribed preventive (prophylactic) antibiotics especially during and after particular types of treatment.

### Things to look out for

At any stage during your treatment it is important that you contact your doctor or the nursing team at the hospital for advice immediately (at any time of the day or night) if you have any problems, if you are feeling unwell or if you experience any of the following:

- a temperature of 38°C or more and/or an episode of uncontrolled shivering
- bleeding or bruising, for example blood in your urine, bowel motions, coughing up blood, bleeding gums or a persistent nose bleed
- nausea or vomiting that prevents you from eating or drinking or taking your normal medications
- diarrhoea, stomach cramps or constipation
- persistent coughing or shortness of breath
- the presence of a new rash, reddening of the skin, itching
- a persistent headache
- a new severe pain or persistent unexplained soreness anywhere
- if you cut or otherwise injure yourself causing excessive bleeding
- if you think you might have had contact with someone with a contagious illness, for example chicken pox, measles, shingles or someone who has had a live vaccine like polio

Don't feel that you are bothering busy people at the hospital. It is in your interest that you stay as well as possible, and presenting earlier may avoid the need to be admitted to hospital. It is very important to deal with any problems that might arise as soon as possible. The sooner they are treated the sooner you will recover.

### Prevention of infection

It is important that you use your common sense when it comes to the prevention of infection. Ask your doctor if you have any questions about this issue. For example, you may wish to go overseas or attend an event or gathering where you think you might be putting yourself at some risk. Your doctor will be able to advise you on the best ways of protecting yourself while living a relatively normal life during this time.

You are at most risk of infection if neutropenic, however, simple precautions to reduce your risk of infection include:

- regular hand washing
- regular mouth care
- avoiding close contact with people with suspected colds, flu and other viruses
- avoiding people who have been in contact with children with chicken pox or measles or other viruses, or children who have had a live vaccine such as polio
- using gloves and face mask when in contact with garden soil, potting mix, compost and grass clippings

- avoiding building dust, such as from major DIY or home renovations
- Only eating food that has been properly cooked and stored

### Treatment of infection

In spite of all these precautions, infections can happen. Infections can be serious and need to be treated with antibiotics as soon as possible.

It is important you do not use treatments to lower your high temperature (i.e. paracetamol) until you are reviewed by your doctor. This could mask an infection which could lead to serious life threatening complications. Do not take aspirin or ibuprofen in any form as this can increase the risk of bleeding if your platelets are low.

Sometimes it is not possible to find the cause of your infection. If you develop a temperature while your white cell count is very low you will be given intravenous antibiotics straightaway. This is to help prevent the spread of infection in the blood. You may also be offered paracetamol to help relieve the symptoms associated with an infection. If the source of the infection is found, the doctors might choose a different antibiotic, one that treats the infection more effectively. If your temperature has not returned to normal within a few days they might decide to use a different antibiotic again, or to add in an anti-fungal drug or an antiviral medication.

You may be feeling quite miserable and unwell if you are febrile. Measures will be taken to limit the infection and to make you as comfortable as possible until it subsides.

### **Blood and platelet transfusions**

People with MDS often need platelet and red blood cell transfusions. If symptoms of anaemia are interfering with your normal daily activities, your doctor may recommend that you have a red blood cell transfusion. Platelet transfusions may be given to treat bleeding (for example a persistent nose bleed), or sometimes during chemotherapy to prevent bleeding.

You do not necessarily need to be admitted to hospital for a red blood cell or platelet transfusion, they are usually given in the clinic or outpatient department of the hospital. Transfusions are very safe and they do not usually cause any serious complications. Nevertheless you will be carefully monitored throughout the transfusion. If you feel hot, cold, and /or shivery or in any way unwell during a transfusion, alert your nurse, 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 do not happen again.

All blood donors and donated blood are carefully screened to minimise the risk of harmful infections being passed on in a transfusion. Careful checks are made both in the blood bank and at the chair-side to ensure that the blood products you are receiving are compatible with your blood type.

## Side effects of repeated transfusions

### **Risk of infection**

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

### **Transfusion reactions**

Careful checks are made both in the blood bank and at the chair-side 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 blood cells present in bags of donated red blood cells and platelets. More recently, these reactions have been dramatically reduced by the use of special filters which remove the white cells at the time of donation.

### **Fluid overload**

Each bag of red blood cells adds nearly 400 ml 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 diuretic medicine may be given to help you pass urine. The nurse will ensure that you are informed when this is necessary 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 red blood cell transfusions can lead to a build up of high levels of iron in the body. Your doctor will be able to tell if this is happening from a simple blood test. Clinical trials are in progress to see if removing excess body iron using a type of drug called a chelating agent will improve patient survival.

### **Growth factors**

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 may be used as medicines to help manage your condition.

Erythropoietin (EPO) is an example of a growth factor which is used to stimulate the production of more red blood cells, and can in some cases reduce the need for frequent blood transfusions. Erythropoietin can cause blood pressure to rise and can increase the risk of blood clots (thrombosis). At this point in time, EPO is not publically funded in NZ for patients with MDS.

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.

Thrombopoietin is a naturally-occurring growth factor that stimulates the production of platelets in the bone marrow. Medicines that mimic the action of thrombopoietin are being tested in MDS in clinical studies, and are not routinely available at present.

Growth factors are given as an injection under the skin (subcutaneous). They do not 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.

## **Complementary therapies**

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

You might consider learning some relaxation techniques such as meditation, yoga or breathing exercises that you can use if feeling anxious or nervous.

Complementary therapies should 'complement' or assist with recommended medical treatment. They are not recommended as an alternative to medical treatment. It is important to realise that no complementary or alternative treatment alone has proven to be effective against MDS.

It is also important to let your doctor or nurse know if you are using any complementary or alternative therapies in case they interfere with the effectiveness of chemotherapy or other treatments or medications you may be having.

## **Nutrition**

A healthy and nutritious diet is important in helping your body to cope with MDS 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 balanced and nutritious diet. This is especially important if you are finding it difficult to eat and/or if you are losing weight.

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 treatments you are having.



## Mouth care

Regular brushing of teeth and examinations by a dentist are important. Keeping your mouth clean, especially if it is sore, helps prevent infection, as does keeping your lips moist with a lip balm or chapstick to avoid cracking. It is important that you report any soreness in your mouth, bleeding gums or if an ulcer or a cold sore develops. Occasionally mouth pain or mouth ulcers may develop as a result of infection e.g. thrush (*Candida albicans*), cold sores (Herpes simplex). These need specific treatment, which will be prescribed by your doctor.

## Fatigue

Feelings of tiredness and even exhaustion are often a side effect of MDS, but can also be a side effect of treatment.

Feeling like you have no energy can be very frustrating, especially if you are used to leading an active and busy life. Try to get plenty of rest but also try to take a little light exercise each day. Getting out into the fresh air and doing some gentle exercise is important for your general feeling of wellbeing and it also may help to give you more energy.

Perhaps you are a member of a gym or sporting club. You might like to ask your doctor about gradually increasing the amount of exercise you do over time and when you might be able to return to your previous way of exercising.

Fatigue may also be a symptom of anaemia. Your blood count will be monitored regularly and you will be given a blood transfusion if you need one. Also, check your platelet count before doing strenuous exercise.

## Palliative care

Palliative care is aimed at relieving any symptoms or pain you might be experiencing as a result of your disease or its treatment, rather than trying to cure or control it. Palliative care is not purely aimed at end of life care, and palliative care nurses or doctors may be involved in your care throughout treatment to help with problematic symptoms, for example pain control or nausea.

If a decision is made to discontinue active treatment for you, there are still many things that can be done to help you to stay as healthy and comfortable as possible for as long as possible. This may involve transfusions, treatment of infections or medicines to reduce symptoms like bleeding or pain.

## Reproductive health

As discussed earlier, MDS is rare in men and women of child-bearing age. However, we have included this section should concerns about fertility be present for you.

### Fertility

Fertility is the ability to produce a child. In males, fertility means having enough healthy sperm to get a female pregnant. In females, fertility is the ability to become pregnant.

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

In women, some types of chemotherapy and radiotherapy can cause varying degrees of damage to the normal functioning of the ovaries. In some cases this leads to menopause (change of life) earlier than expected. In men sperm production can be impaired for a while but the production of new sperm may become normal again in the future.

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

#### Protecting your fertility - Men

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

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

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

### Protecting your fertility - Women

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

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

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

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

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

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

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

### Early menopause

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

Hormone changes can lead to many of the classic symptoms of menopause including menstrual changes, hot flushes, sweating, dry skin, vaginal dryness and itchiness, headache and other aches and pains. Some women experience decreased sexual drive, anxiety and even depressive symptoms during this time.

It is important that you discuss any changes to your periods with your doctor or nurse. He or she may be able to advise you or refer you to a specialist doctor (a gynaecologist) or clinic that can suggest appropriate steps to take to reduce your symptoms.

### Menstruation

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

### Body image, sexuality and sexual activity

The diagnosis and treatment of MDS may have had some impact on how you feel about yourself as a man or a woman and as a 'sexual being'. Hair loss, skin changes and fatigue can all interfere with feeling attractive.

During treatment you may experience a decrease in libido, which is your body's sexual urge or desire, sometimes without there being any obvious reason. It may take some time for things to return to 'normal'. It is perfectly reasonable and safe to have sex while you are on treatment or shortly afterwards, but there are some precautions you need to take. Men may sometimes experience some degree of erectile dysfunction during treatment and in the stages of recovery. If this is an issue for you, please discuss this with your doctor so any help can be initiated.



It is usually recommended that you or your partner do not attempt to become pregnant while undergoing treatment, as some of the treatments given might harm the developing baby. As such, you need to ensure that you or your partner uses a suitable form of contraception. Condoms (with a spermicidal gel) offer good contraceptive protection as well as protection against infection or irritation. Your partner may be worried that sex might in some way harm you. This is not likely as long as your partner is free from any infections and the sex is relatively gentle. It is important to be extra careful when your white cell count or platelet levels are low as you may be more prone to infection and bleeding/spotting. Finally, if you are experiencing vaginal dryness, a lubricant can be helpful. This will help prevent irritation. Using a condom is also important to protect your partner from chemotherapy drugs that can be excreted in body fluids in the first few days after they are administered.

If you have any questions or concerns regarding sexual activity and contraception don't hesitate to discuss these with your doctor or nurse, or ask for a referral to a doctor or health professional who specialises in sexual issues.



## Social and emotional effects

People cope with being diagnosed with MDS in different ways, and there is no right or wrong or standard reaction. For some people, it can trigger any number of emotional responses ranging from denial to devastation. It is not uncommon to feel angry, helpless and confused. Naturally people fear for their own lives or that of a loved one.

It is worth remembering that information can often help to take away the fear of the unknown. It is a good idea for you and your family to speak directly to your doctor regarding any questions you might have about your disease or treatment. It can also be helpful to talk to other health professionals including social workers or nurses who have been specially educated to take care of people with blood conditions. Some people find it useful to talk with other patients and family members who understand the complexity of feelings and the kinds of issues that come up for people living with blood cancers and conditions.

In some areas there may be Leukaemia & Blood Cancer New Zealand education and support group meetings, and there is also an opportunity to connect with others online via [www.leukaemia.org.nz](http://www.leukaemia.org.nz) or our Facebook page: [www.facebook.com/LBFNZ](https://www.facebook.com/LBFNZ).

Many people are concerned about the social and financial impact of the diagnosis and treatment on their families. Normal family routines are often disrupted and other members of the family may suddenly have to fulfil roles they are not familiar with, for example, cooking, cleaning, and taking care of children. The social worker attached to your treatment centre is available for you to talk through and discuss these issues with you and your family.

If you have a psychological or psychiatric condition, please inform your doctor and don't hesitate to request additional support from a mental health professional.

There is a variety of assistance available to help ease the emotional and financial strain created by a diagnosis of a blood cancer or condition. In some cases travel assistance may be available. Support Services staff at Leukaemia & Blood Cancer New Zealand are available to provide you and your family with information and support to help you cope during this time.

Contact details for Leukaemia & Blood Cancer New Zealand are provided on the back of this booklet.



## Useful internet addresses

The value of the internet is widely recognised; however, not all the information available may be accurate and up to date. For this reason, we have selected some of the key sites that people with MDS might find useful.

With the exception of our own websites, Leukaemia & Blood Cancer New Zealand do not maintain these listed sites. We have only suggested sites we believe may offer credible and responsible information, but we cannot guarantee the information on them is correct, up to date or evidence based medical information.

### **Leukaemia & Blood Cancer New Zealand**

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

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

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

### **Leukaemia Foundation of Australia**

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

### **American Cancer Society**

[www.cancer.org](http://www.cancer.org)

### **Aplastic Anaemia & MDS International Foundation (US)**

[www.aamds.org](http://www.aamds.org)

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

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

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

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

### **Myelodysplastic Syndromes Foundation (US)**

[www.mds-foundation.org](http://www.mds-foundation.org)

### **MacMillan Cancer Support (UK)**

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

### **National Cancer Institute (US)**

[www.cancer.gov/cancerinfo](http://www.cancer.gov/cancerinfo)

## Glossary of terms

### **Alopecia**

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

### **Anaemia**

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

### **Antibodies**

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

### **Antiemetic**

Drug which prevents or reduces feelings of sickness.

### **Blasts**

Immature stem cells, normally making up no more than 5% of cells in the bone marrow. Blast cells are not normally found in healthy peripheral blood.

### **Bone marrow**

The tissue found at the centre of many flat or big bones of the body. The bone marrow contains stem cells from which all blood cells are made. Active or red bone marrow contains stem cells from which all blood cells are made and in adults 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.

### **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, 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 cancerous, these 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.

### **Cells**

Small particulate components of the human body consisting of fluid cytoplasm with or without a central nucleus, enclosed in a membrane. In blood, cells can be red cells, white cells or platelets.

### **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 40,000 genes on 23 different chromosomes. The chromosomes are contained within the nucleus of a cell.

### **Cure**

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

### **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.

### **High dose therapy**

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

### **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 they can cause very high abnormal white cell counts.

**Lymphocytes**

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 of 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.

**Resistant or refractory disease**

This means that the disease is not responding to treatment.

**Spleen**

The spleen is found high in the abdomen on the left-hand side and cannot normally be felt on examination unless it is enlarged. It is an organ that is part of the blood system and is a specialized collection of lymphoid and haematopoietic tissue. It plays a minor role in the immune system and contributes to the destruction of red blood cells, white blood cells and platelets at the end of their lifespan. It is often enlarged in diseases of the blood - this is known as hypersplenism.

**Splenomegaly**

Enlargement of the 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 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.

**Tumour**

An abnormal mass of cells which may be non-malignant (benign) or malignant (cancerous).

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

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

- |  |  |
|--|--|
| <input type="checkbox"/> Dictionary of Terms   | <input type="checkbox"/> Acute Lymphoblastic Leukaemia in Adults   |
| <input type="checkbox"/> Haematology Patient Diary                                   | <input type="checkbox"/> Acute Lymphoblastic Leukaemia in Children |
| <input type="checkbox"/> Clinical Trials   | <input type="checkbox"/> Acute Myeloid Leukaemia                   |
| <input type="checkbox"/> Autologous Stem Cell Transplants                            | <input type="checkbox"/> Chronic Lymphocytic Leukaemia             |
| <input type="checkbox"/> Allogeneic Stem Cell Transplants                            | <input type="checkbox"/> Chronic Myeloid Leukaemia                 |
| <input type="checkbox"/> Myeloproliferative Disorders                                | <input type="checkbox"/> Hodgkin Lymphoma                          |
| <input type="checkbox"/> Myelodysplastic Syndromes                                   | <input type="checkbox"/> Non-Hodgkin Lymphoma                      |
| <input type="checkbox"/> Myeloma   | <input type="checkbox"/> Haemochromatosis                          |
| <input type="checkbox"/> My Guide to Blood Cancer - for adolescents and young adults |  |

**Or information on:**

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

**Newsletters:**

- |   |  |
|---|--|
| <input type="checkbox"/> LifeBlood      | <input type="checkbox"/> Leukaemia Today |
| <input type="checkbox"/> Lymphoma Today | <input type="checkbox"/> Myeloma Today   |

Name: \_\_\_\_\_

Address: \_\_\_\_\_

Postcode: \_\_\_\_\_ Phone: \_\_\_\_\_

Email: \_\_\_\_\_

Send to: Leukaemia & Blood Cancer New Zealand  
 PO Box 99182, Newmarket, Auckland 1149  
 Phone: 09 638 3556 or 0800 15 10 15  
 Email: [info@leukaemia.org.nz](mailto:info@leukaemia.org.nz)

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

## Notes

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**Myelodysplastic Syndromes (MDS)**

We hope that you found this information booklet useful. We are interested in what you thought of the booklet – whether you found it helpful or not. If you would like to give us your feedback, please fill out this questionnaire and send it to Leukaemia & Blood Cancer New Zealand, at the address at the bottom of the following page.

**1. Did you find this booklet helpful?**

Yes     No

Comments \_\_\_\_\_

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**2. Did you find this booklet easy to understand?**

Yes     No

Comments \_\_\_\_\_

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**3. Where did you get this booklet from?**


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

Yes     No

If yes, what were they?

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


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


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**7. Any other comments?**


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Thank you for helping us review this booklet. We will record your feedback and consider it when this booklet is reviewed for the next edition.

Please return to: Leukaemia & Blood Cancer New Zealand  
PO Box 99182 Newmarket, Auckland 1149



## Important information for haematology patients

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

- a temperature of 38°C or over and / or an episode of shivering
- bleeding or bruising, for example blood in your urine, faeces, sputum, bleeding gums or a persistent nose bleed
- nausea or vomiting that prevents you from eating or drinking or taking your normal medications
- diarrhoea, stomach cramps or constipation
- coughing or shortness of breath
- the presence of a new rash, reddening of the skin, itching
- a persistent headache
- a new pain or soreness anywhere
- if you cut or otherwise injure yourself
- if you notice pain, swelling, redness or pus anywhere on your body





our mission is to care, our vision is to cure

**Freephone** 0800 15 10 15  
**Telephone** 09 638 3556  
**Facsimile** 09 638 3557  
**Email** [info@leukaemia.org.nz](mailto:info@leukaemia.org.nz)

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**[leukaemia.org.nz](http://leukaemia.org.nz)**

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