Myeloproliferative Neoplasms (MPNs)

A guide for patients & families

UHN Princess Margaret Cancer Centre

What's inside

In this booklet you will find information on your care at Princess Margaret and on MPNs.

This booklet covers the basics on MPNs. This includes:

- What MPNs are
- What symptoms to expect
- Living with an MPN
- Treatments you may be offered
- Support groups and where to get more information

You will also find information on what to expect in your care at Princess Margaret and who to contact if you have questions or concerns.

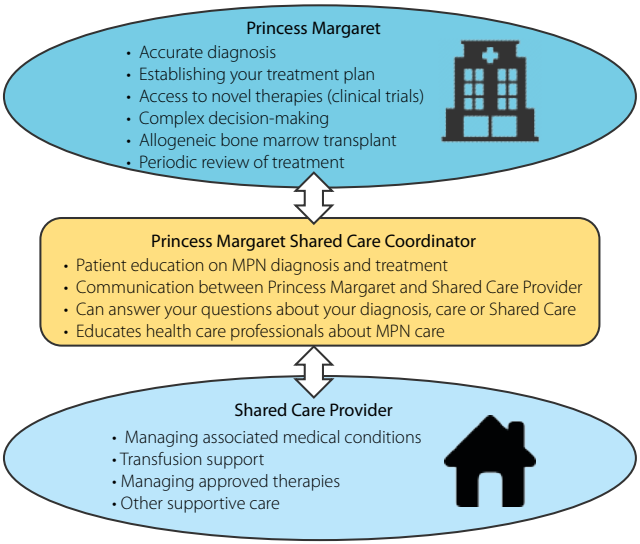
If there is something you do not understand or is not answered, please talk to your physician or anyone in your MPN health care team.

Contents

Shared Care

Princess Margaret Cancer Centre is a world-renowned centre with leading experts in a variety of cancer fields. This allows us to provide some specialist care that may not be available closer to your home.

The Elizabeth and Tony Comper Myeloproliferative Neoplasm Program provides MPN diagnosis, therapy and education recommendations, and involves your referring healthcare practitioner throughout your MPN journey. This philosophy is known as ***Shared Care***.



Shared Care is a model which allows you to maintain contact with Princess Margaret Cancer Centre, without needing to travel very often for care. You will receive some of your MPN care at Princess Margaret (how much vary depending on your disease and your treatment) while your Shared Care provider (primary haematologist, oncologist, internal medicine specialist or family physician) continues to monitor you closer to home. Shared care provides assurance that you ***are always cared for***.

An Introduction to MPNs

What does it mean?

Myeloproliferative Neoplasms (MPNs)

Myelo – to do with cells from the bone marrow

Proliferative – an overproduction

Neoplasm – an abnormal growth of cells, in this case blood cells

Myeloproliferative Neoplasms (MPNs) are a group of rare disorders that affect the production of some blood cells with specific functions (such as red blood cells and platelets).

Blood cells are produced from multipurpose stem cells in the bone marrow. The change in stem cells results in abnormal production of different types of blood cells.

MPNs are not usually hereditary, meaning parents do not pass it on to their children. MPN has been found to run in a small number of families but this is very rare.

This booklet provides information about 3 of the main MPN disorders:

- Polycythemia vera (PV)
- Essential thrombocythemia (ET)
- Myelofibrosis (MF).

Polcythemia vera (PV)

Polycythemia vera (PV) is a disorder characterized by an **overproduction of red blood cells** in the bone marrow.

Red blood cells transport oxygen around the body. Too many red blood cells results in blood that is thicker than normal, which can cause problems such as blood clots or bleeding. White blood cells (which fight infection) and platelets (which help blood to clot) can also be increased.

Essential thrombocythemia (ET)

Essential thrombocythemia (ET) is characterized by an **overproduction of platelets** in the bone marrow. Platelets are blood cells that help blood to clot but too many can cause blood clots or excessive bleeding.

Myelofibrosis (MF)

Myelofibrosis (MF) is less common than ET or PV. **Scar tissue forms in the bone marrow** in patients with MF. This scar tissue interferes with the production of normal, healthy blood cells that normally happens in the bone marrow. Myelofibrosis can occur on its own (primary MF) or it can develop in individuals who have had PV or ET for a number of years.

MDS/MPN overlap

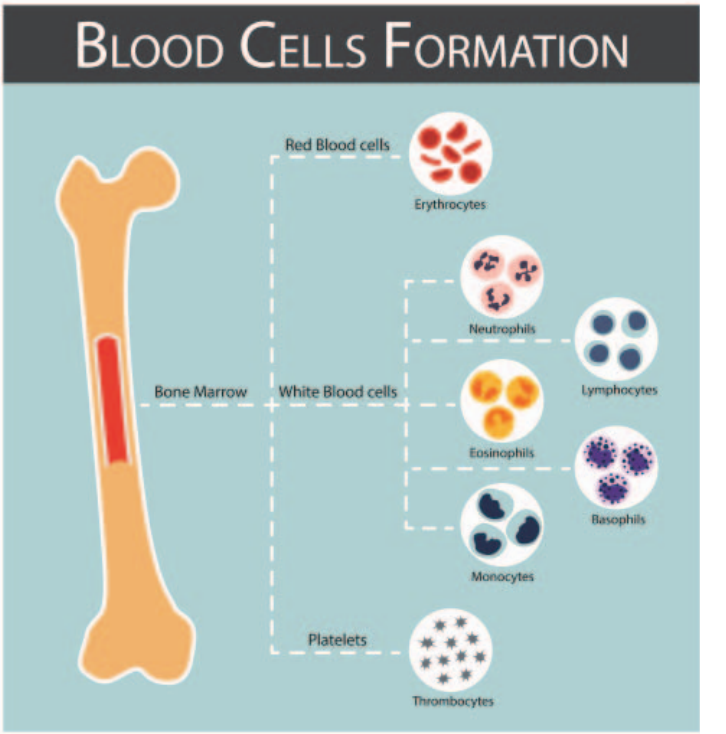
MPNs can sometimes overlap with myelodysplastic syndrome (MDS). In MDS stem cells in the bone marrow do not mature or they produce dysplastic (abnormal) blood cells. Patients with blood cells that show proliferative (overproduction, like an MPN) and dysplastic (abnormal, like MDS) characteristics are diagnosed with MDS/MPN.

How do MPNs affect my blood?

MPNs affect the production of hematopoietic stem cells

Stem cells are the beginning of every cell, and are programmed to grow into specialized mature cells of many types, such as blood, muscle or brain cells.

Stem cells that mature into red blood cells, white blood cells and platelets are known as hematopoietic stem cells. Your bone marrow is responsible for producing hematopoietic stem cells. After they are mature, blood cells are released from the bone marrow into the blood circulation.



Types of blood cells made by the bone marrow

Hematopoietic stem cells in the bone marrow mature into…

Red blood cells - help to transport oxygen to cells via haemoglobin. Their circulating life span is about 120 days.

Platelets - help to regulate blood clotting in the body (serve as the plug when you get a cut). Circulating life span 10 days.

White blood cells - infection fighting cells (neutrophils, basophils, eosinophils, monocytes, lymphocytes). Circulating life span 7-21 days.

What do my blood counts mean?

The result of a complete blood count (CBC) provides a haematologist with a picture of how your bone marrow is working. Some of the main values that are reviewed are:

Normal values

Hemoglobin (Hb)

Female 123 – 157 g/L

Male 140 – 174 g/L

White blood cells (WBC)

4 – 10 x 10^9/L

(4 –10 billion per litre)

Hematocrit (HCT)

Female 0.370 – 0.460

Male 0.420 – 0.520

Platelet (Plts)

150 – 400 x 10^9 /L (150–400 billion per litre)

How do MPNs affect blood counts?

An increase in hemoglobin (Hb) and/or haematocrit (HCT) significantly over and above the normal values consistently for both men and women can indicate polycythemia vera.

An increase in platelet count (Plts) significantly over and above the normal values consistently for both men and women can indicate essential thrombocythemia (ET).

Blood results that show low hemoglobin (Hb), red blood cells, haematocrit (HCT) and immature blood cells, may indicate myelofibrosis if splenomegaly (big spleen) is also present.

Blood work is not the only indication for diagnosis. Signs and symptoms of ET/PV & MF are important — these will be discussed further in this booklet.

Mutations in MPN

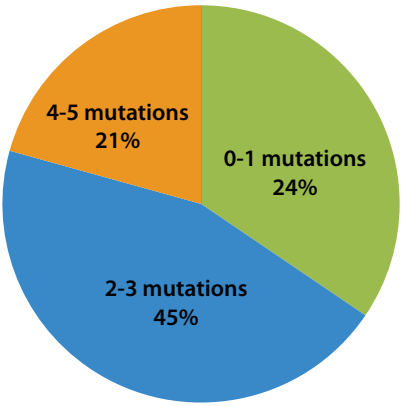
The exact causes of MPNs are not fully understood. However, most MPN patients have one of the commonly seen mutations in certain proteins. Mutations are changes in the protein. These proteins help the hematopoietic stem cells in the bone marrow produce healthy blood cells. The mutations make the hematopoietic stem cells in the bone marrow less able to produce normal blood cells.

Most MPN patients have more than one mutation.

MPNs are complex diseases and most patients have multiple mutations.

This almost always includes a JAK2, CALR or MPL mutation (read more on this below).

Average number of mutations in MPN patients



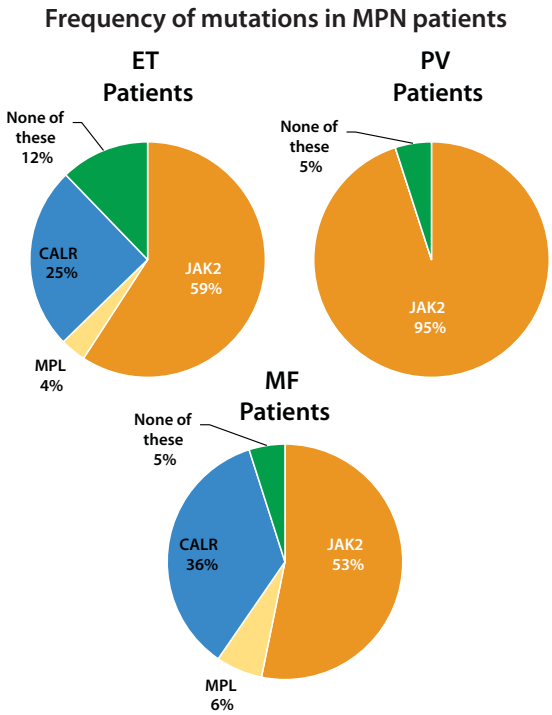
(unpublished data from MPN Program, Princess Margaret)

Key MPN mutations

JAK2: many patients with an MPN have a mutation in the JAK2 protein, which helps regulate blood cell production.

CALR and MPL: a smaller number of MPN patients have a mutation in the calreticulin (CALR) protein or the myeloproliferative (MPL) protein — these proteins also help regulate blood cell production.

The diagrams below show how common these mutations are in MPN patients.



Adapted from data in Klampfl et al., N Engl J Med 2013;369:2379-90.)

Almost all PV patients and most ET and MF patients have one of these mutations. For this reason, all MPN patients are tested for the JAK2 mutation and may also be tested for CALR or MPL.

Research on MPN mutations

JAK2 and CALR are the most frequent mutations seen but mutations in many other proteins are found as well. However, we don't understand the significance of most of these.

Important: the significance of many mutations (apart from JAK2 and CALR) is currently unknown. These mutations are the subject of ongoing clinical research.

AGILE research study

To increase our understanding of mutations in MPNs, all MPN patients at Princess Margaret are invited to take part in a clinical research study called "AGILE". AGILE tests for mutations in many proteins. It is a research study and not a standard of care test (unlike testing for JAK2 for example). You will be given more information about this at your appointment and taking part in this study is voluntary.

We don't how or if many of these mutations affect your MPN, so AGILE study results are not expected to directly affect the decisions your physician makes about your care, but taking part in studies like this helps us improve understanding and ultimately treatment of MPNs.

How does having an MPN affect my heart?

Your heart is essential for providing your cells with oxygen and nutrients by pumping blood to all parts of your body.

Risk of blood clots

PV and ET patients have a higher risk of developing blood clots (thrombosis).

In ET and PV, the high number of red blood cells and platelets can fill the arteries (blood vessels that carry blood away from the heart) or veins (blood vessels that return blood to the heart).

This can cause a clot (thrombosis) and can block blood flow. This can lead to stroke, deep vein thrombosis, or heart attack.

High cholesterol

Another factor that can affect the heart is the development of atherosclerosis (plaque build-up in the arteries).

You doctor may put you on a 'statin' to help control cholesterol levels to decrease the chance of a heart attack or stroke.

High blood pressure

High blood pressure (hypertension) can also cause damage to blood vessels and the heart.

Your doctor may prescribe blood pressure medication to keep it within a normal range that is safe for you.

Diabetes

Diabetes control is important as well if you have an MPN. Diabetes can affect blood vessels and the heart if it is not well controlled.

Smoking

Smoking also impacts the circulation of blood, and increases your risk for heart disease and stroke. Smoking can contribute to plaque build-up in the arteries, reduce oxygen levels in the blood and increases your blood pressure.

For tips on how to quit smoking and resources available to help you see p.39..

Essential Thrombocythemia

What does it mean?

Pronounced: throm-boe-sie-thee-me-uh

Thrombo – identifies it as a condition involving platelets (thrombocytes), which are involved in blood clotting.

Cythemia – identifies it as a condition involving cells in the blood

What is it?

Essential thrombocythemia (ET), sometimes referred to as primary thrombocytosis, is a rare chronic bone marrow disorder characterized by the **overproduction of blood platelets** in the bone marrow. Platelets are involved in the clotting of blood and the increased number of platelets in the blood can put patients at increased risk of blood clots or excessive bleeding.

ET is mainly diagnosed in older individuals over 60 years of age, but can be diagnosed in patients less than 40 years of age. ET is a rare disorder - approximately 1,700 Canadians have ET.

About 20-25% of patients are diagnosed after a cardiac or neurological event. And 20-25% of those diagnosed may develop a blood clot (thrombotic event).

Symptoms

Many ET patients have no symptoms and patients are commonly diagnosed after a routine check-up. Others may experience some of the common ET symptoms below.

Common ET symptoms

- Nose bleeds
- Bruising
- Headaches
- Burning, redness and pain in the fingertips or toes
- Swelling of the spleen
- Fatigue
- Dizziness or light-headedness
- Vision disturbances or silent migraines

Cause

The causes of ET are not fully understood. However, research has shown that about 50% of ET patients have a mutation in the JAK2 protein, which helps regulate blood cell production.

Other protein mutations have also been implicated. About 20-25% of ET patients have a calreticulin (CALR) protein mutation and fewer than 5% of ET patients have a MPL protein mutation. These proteins also contribute to normal blood cell production.

Living with ET

Prognosis

The life expectancy of patients with ET depends on whether the individual has low, medium or high risk disease. Patients with low risk disease and who do not suffer from another disease have a normal to slightly reduced life expectancy. Prognosis depends on many factors including age, cardiovascular illnesses and complications.

The most common complications and causes of death in ET patients are related to increased platelet count such as bleeding and blood clots (thrombosis)

Disease progression

For most patients diagnosed with ET their disease remains stable over time. In a small minority of patients with ET their disease progresses to myelofibrosis (known as post-ET myelofibrosis) and in rare cases to acute myeloid leukemia, both of which can be life threatening.

Myelofibrosis (MF) is an MPN disorder where scarring develops in the bone marrow, interfering with normal blood cell production. About 3-5% of patients will ET progress to MF at 10 years after diagnosis. You can read more about myelofibrosis on p. 20.

Acute myeloid leukemia (AML) is a type of leukemia that progresses rapidly. Post-ET AML is an aggressive disease and is often resistant to treatment. About 2-5% of patients will ET progress to AML at 10-15 years after diagnosis.

Treatment

There is currently no cure for ET. The main **goal of treatment is to reduce the platelet count** to reduce the risk of complications, such as bleeding and abnormal blood clotting (thrombosis).

Because patients with ET are at a higher risk of developing a cardiovascular complication such as blood clots, stroke or heart attack, treatment also includes managing cardiovascular risk factors — see p11 for more information.

The most common treatment is **daily low-dose aspirin**. Other therapy options include hydroxyurea, anagrelide, interferon and busulphan — these treatments can reduce the platelet count, as well as other counts.

The role of JAK inhibitors in the treatment of ET remains investigational with other therapies

Read more about treatments on p.25

Polycythemia Vera

What does it mean?

Pronounced: pol-e-sigh-thee-me-uh vayr-uh

Cythemia – identifies it as a condition involving cells in the blood.

Poly – means many. Too many red blood cells are being produced.

What is it?

Polycythemia vera (PV) is a rare chronic disease of the bone marrow in which **too many red blood cells are produced.** This makes the blood thicker than normal and increases the risk of blood clots and bleeding. Clots can block blood flow through the arteries and veins, which can lead to complications such as heart attack or stroke.

Other types of blood cell also produced in the bone marrow can also be increased. These include white blood cells (which fight infection) or platelets (which help blood to clot). With the bone marrow not functioning properly, the spleen often compensates by producing red blood cells, which causes it to enlarge in most PV patients.

Most people diagnosed with PV are 60 years old or more.

Symptoms

In some patients, PV is diagnosed after blood work or a physical examination during a routine doctor's visit. Some common PV symptoms are listed below.

Common PV symptoms

- Headaches
- Blurred vision
- Weakness
- Dizziness
- Fatigue (tiredness)
- Bleeding or clotting
- Bone pain
- Skin itching (pruritus) – especially after showering
- Feeling full quickly – caused by an enlarged spleen pressing against the stomach
- Redness and burning pain in the feet

The kind and severity of symptoms experienced varies a lot between patients. The most common symptom experienced by patients with PV is fatigue.

Cause

The causes of PV are not known. However, nearly 95% of people with PV have a mutation in the JAK2 protein, which helps regulate blood cell production. This mutation leads to abnormal blood cell production.

Living with PV

Prognosis

PV is a chronic disease and people who have well controlled disease, do not suffer from other diseases (especially myelofibrosis or leukemia) have a normal to slightly reduced life expectancy.

Prognosis depends on many factors including age, other cardiovascular illnesses, and PV complications. The most common complications and causes of death in PV patients are related to an increased risk of blood clotting (thrombosis), such as heart attacks and stroke. About 45% of PV-related deaths are associated with cardiovascular disease. The goal of PV treatment is to reduce the risk of these complications.

Disease progression

In a minority of patients PV can transform into a more aggressive form.

Myelofibrosis: At 10-15 years after PV diagnosis about 15% of people with PV progress to myelofibrosis, known as post-PV myelofibrosis (post-PV MF). MF is an MPN disorder where scarring develops in the bone marrow, interfering with normal blood cell production. Transformation of PV to MF results in a reduced life expectancy.

Acute myeloid leukemia: Rarely, some patients with PV transform to acute myeloid leukemia (AML). Post-PV AML is an aggressive disease and is often resistant to treatment.

Treatment

There is currently no cure for PV. The goal of treatment is to reduce the number of blood cells and help to maintain normal blood counts as well as lower the risk of blood clots and other complications. Treatment also includes managing cardiovascular risk factors — see p.11 for more information.

Treatment options are aimed at reducing the number of blood cells and the risk of blood clots. They include:

- Phlebotomy (blood letting)
- Hydroxurea
- Interferon
- Anagrelide
- Busulphan
- Aspirin

Read more about treatments on p.25

Patients at low risk for complications are primarily treated with phlebotomy and low-dose aspirin.

The JAK inhibitor Jakavi® (ruxolitinib) has been approved to treat some PV patients. Your physician may consider Jakavi® if your disease is not well controlled with conventional cytoreductive therapy.

Myelofibrosis

What does it mean?

Pronounced: my-ah-lo-fi-bro-sis

myelo – means that the disease affects blood cells in the bone marrow.

fibrosis – relates to an excessive scarring of tissue in the bone marrow that impairs its ability to produce normal blood cells..

What is it?

Myelofibrosis (MF) is the least common MPN, but the most debilitating. Scar tissue forms in the bone marrow, impairing its ability to produce normal blood cells. This leads to severe anemia (low red blood cell count / low hemoglobin), weakness and fatigue.

Cause

The causes of MF are not fully understood. However, research has shown that about 50% of MF patients have a mutation in the JAK2 protein, which helps regulate blood cell production. Research has shown that some MF patients have mutations in other proteins, including CLAR and MPL, which are also involved in normal blood cell production.

MF may occur on its own or as a result of other types of MPN.

The types of MF are:

- ***Primary MF:*** myelofibrosis that occurs on its own
- ***Post-PV MF:*** post-polycythemia vera myelofibrosis
- ***Post-ET MF:*** post-essential thromboycthemia myelofibrosis (MF)

Symptoms

Symptoms of MF are different for every person. Early in the disease patients may have very few or no symptoms. Symptoms of MF are usually related to low blood counts, enlarged spleen or symptoms such as fever, night sweats and weight loss.

As the bone marrow becomes less able to create normal blood cells, blood cell production may move to the spleen and liver, causing them to swell and enlarge.

Common PV symptoms

- Fatigue
- Fever
- Night sweats
- Bone & muscle pain
- Weight loss
- Abdominal pain
- Itchiness
- Pain under the left ribs
- Feeling full quickly – caused by an enlarged spleen pressing against the stomach
- Easy bruising or bleeding

Living with MF

Prognosis

The prognosis of MF is different for every patient. People in a good prognostic group can live for many years without having major symptoms; those with a poor prognosis may progress more quickly. There are many factors that determine prognosis, including age, blood cell counts and symptoms such as night sweats, fever and weight loss.

Disease progression

In a small number of patients with MF (20-30%) progress to a form of leukemia called acute myeloid leukemia (AML). AML arising from MF is an aggressive disease and current treatments are not very effective to achieve remission.

How is MF risk calculated?

Several tools are used to calculate the progression of an MPN over a period of time. One is the International Prognosticator Scoring System (IPSS); another is the Dynamic International Scoring System (DIPSS). Physicians also use a system called DIPSS Plus, which is more extensive than DIPSS.

There are also several other tools that consider mutations when calculating MF risk. These tools are still under development and active research.

These scoring systems take into account risk factors such as: symptoms, physical assessment (such as enlarged spleen), blood work abnormalities, bone marrow biopsy results and mutations.

Your doctor can go into more details of this scoring system during your visit.

Treatments

Each patient with myelofibrosis is different. Treatment options range from wait and carefully watch to intensive treatment options such as allogeneic bone marrow transplantation.

Jakavi®

Jakavi® (also called ruxolitinib) is the first drug approved specifically to treat MF patients. Jakavi® reduces the size of the spleen in many MF patients as well as reducing some of the other symptoms of MF (night sweats, weight loss, fever, pruritis).

Allogeneic stem cell transplant (bone marrow transplant)

Allogeneic stem cell transplant is the only therapy that can cure MF. However it is not recommended for patients with lower risk disease. Your physician may recommend a transplant if you have higher risk disease, are experiencing severe constitutional symptoms (e.g. enlarged spleen, night sweats) that are not controlled with other therapies, or if your disease is progressing. See p.25.

Read more about treatments on p.25

Other treatments

Different therapies are used to treat specific symptoms of MF. Below are some common therapies used to treat specific symptoms of MF

Anemia - (low red blood cell count / low hemoglobin) may be treated with:

- Blood transfusions – increase the number of red blood cells in the body
- Corticosteroids
- Androgens (such as danazol)
- Erythropoiesis stimulating agents (ESAs)

These stimulate the body to make red blood cells

An enlarged spleen (splenomegaly) may be treated with:

- Jakavi® (also called ruxolitinib)
- Hydroxyurea

These stimulate the body to make red blood cells

- Interferon
- Radiation or splenectomy (removal of the spleen) may be considered in severe cases

Clinical Trials

Although ruxolitinib is approved for treatment of MF, this treatment has some limitations. For example, ruxolitinib is not suitable for some patients with a low platelet count or anemia (low red blood cells / low hemoglobin) and some patients may experience side effects.

Clinical trials are ongoing to find other therapies for MF that do not have these disadvantages. Your physician may recommend that a clinical trial may be suitable for you. You can find out more about clinical trials on p.29..

Treatments for MPNs

Although there is no cure for MPNs, there are some treatment options which can help manage your symptoms.

Baby Aspirin – helps to thin your blood if you have elevated hemoglobin or platelets. This helps to prevent blood clots (thrombosis).

Phlebotomy – is removal of a set amount of blood to control hematocrit and hemoglobin levels. It is primarily for patients diagnosed with PV.

Hydroxyurea – helps to reduce your white blood cell count and neutrophil count. It also affects your hemoglobin and platelet count.

These 3 treatments (above) try to prevent clots from forming (thrombosis) – to reduce the risk of a cardiac event (e.g. stroke, deep vein thrombosis)

Interferon alpha / pegylated interferon alpha – used if other agents fail to control blood counts. This treatment requires long-term administration to potentially see a reduction in counts and some symptoms. Major side effects include flu-like symptoms, depression and fatigue.

Anagrelide – helps to reduce platelet count only, and is only recommended if hydroxyurea therapy fails. This treatment does not help with splenomegaly and is not recommended for patients with heart conditions.

Busulphan – is a chemotherapy drug which can reduce high red blood cell count in PV patients. It is only used as a last line of therapy due to its potential to increase leukemia risk.

JAK inhibitors – primarily used in MF and most recently in PV. JAK inhibitors help to reduce symptoms and reduce the size of an enlarged spleen. Blood counts may be affected and may require adjustment of your dose or stopping the drug. Currently the only approved JAK inhibitor is ruxolitinib (Jakavi®) but several others are in clinical trials.

Blood transfusions – replace hemoglobin (red blood cells) or platelets. In some patients with MF, the hemoglobin (red blood cell) and platelet count can be too low, and can impact daily activity. Low platelets can also increase the risk of complications (such as bleeding).

Allogeneic Hematopoietic Stem Cell Transplant (HSCT)

Also known as a bone marrow transplant. HSCT is the only curative therapy for MF and is suitable for some patients. It is a medical procedure to transplant the stem cells of a 'matched' donor (related or unrelated to the patient) to the MF patient in order to help the bone marrow function properly and produce healthy hematopoetic stem cells. See p.27 for more information.

All of these interventions require more discussion with your haematologist to see which treatment is best for you.

Side effects

If you experience unpleasant side effects from your medication it is important to tell your doctor. They can make suggestions for medication or other changes to help get the side effects under control.

Drug Reimbursement

Some of the medications prescribed to you can be costly, even if you have private insurance coverage. At Princess Margaret Cancer Centre, we have Medication Reimbursement Specialists (MRS) that can help apply for additional drug coverage.

The MRS will consider your financial status and help find the most appropriate coverage for you. Call 416-946-2830 if you have any questions about applying for additional coverage.

Hematopoeitic Stem Cell Transplantation (HSCT)

What is HSCT?

Hematopoietic stem cell transplantation (HSCT) is also known as a **bone marrow transplant**. It involves transplanting hematopoietic stem cells from a healthy donor to the patient.

HSCT: why and when?

HSCT is the only therapy that can cure MF but because the procedure carries risks, it is only suitable for patients with higher risk disease.

What does HSCT involve?

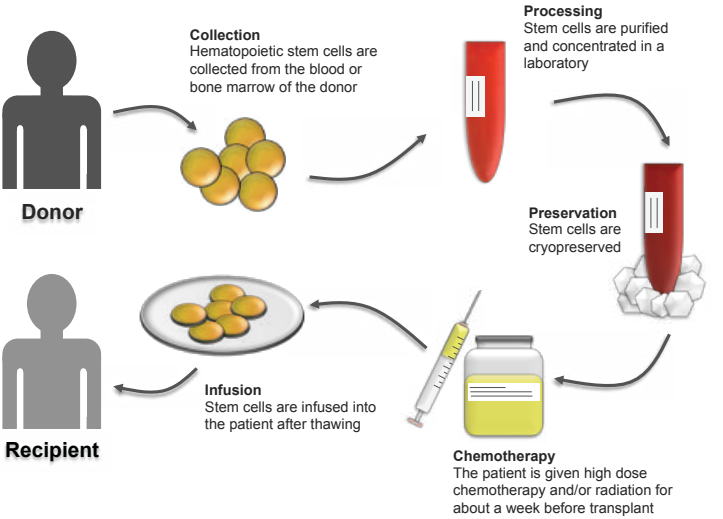
HSCT involves using the stem cells from another donor (related or unrelated) and transplanting them into the recipient after they have undergone chemotherapy (with or without radiation).

The purpose of the chemo/radiation is to prepare the marrow to receive the donor cells and for the cells to reproduce and multiply (proliferate). After a successful transplant, blood counts will gradually improve MF symptoms will gradually reduce.

HSCT involves at least an inpatient stay in hospital for transplantation, then a commitment of follow up appointments to monitor your progress.

Considering HSCT

There are certain risks involved when considering HSCT and this procedure is not successful in all patients. It should be discussed with a member of the HSCT team in order to make an informed decision.


Donor
Collection
Hematopoietic stem cells are collected from the blood or bone marrow of the donor
Processing
Stem cells are purified and concentrated in a laboratory
Preservation
Stem cells are cryopreserved
Chemotherapy
The patient is given high dose chemotherapy and/or radiation for about a week before transplant
Infusion
Stem cells are infused into the patient after thawing
Recipient

Clinical Trials

What are clinical trials?

A clinical trial is a type of research study that is done to help learn about new ways to prevent, diagnose or treat a disease. This type of research involves human beings who choose to participate.

There are two types of clinical trial:

Observational – participants are observed by researchers who monitor their outcomes

Interventional – participants are assigned a treatment (for example a drug) by researchers who monitor their outcomes

Approval of clinical trials

All clinical trials must be approved by a research ethics board before any patient is approached about the study.

The ethical and legal codes that govern medicine also apply to clinical trials.

A research ethics board is a group of individuals with different professional backgrounds that review all clinical trials to make sure that patients' rights are protected.

Why are clinical trials done?

To improve medicine

Clinical trials are one of the last stages of cancer research. Before a clinical trial is started, a lot of promising research has already occurred. Research that shows promise must first be tested in a clinical trial before it can be used in everyday medicine, or as the standard of care.

To provide patients with another option

For patients, clinical trials offer one more choice in their medical care.

Sometimes, patients who participate in clinical trials will have

access to treatments and procedures that otherwise would not be available. However, it is important to understand what is involved in a clinical trial before deciding to participate.

Deciding whether to participate

Clinical trials are important in developing new therapies to treat MPNs.

> Deciding not to participate in a clinical trial will **never** affect your care

It is important that you carefully consider whether to participate in a clinical trial and discuss this decision with your family and doctor. Patients who decide to participate can always change their mind.

It is up to you and your doctor to decide what trial is best for you.

Every clinical trial has strict eligibility criteria that determine which patients can participate in the trial. There are also some factors that may exclude you from a particular clinical trial.

I want to know more about clinical trials…

About clinical trials

There are many resources available to learn about MPN clinical trials. Here are some of them:

Your doctor

Princess Margaret Patient and Family Library: A guide on clinical trials and how they work is available called "Clinical Trials at PMH".

Useful websites:

- Canadian MPN Group: this charitable organization provides a regularly updated list of clinical trials in Canada, which includes details on the trials and which centres are participating. http://www.mpncanada.com
- Ontario Cancer Trials Website: this website has database of

cancer clinical trials in Ontario http://ontariocancertrials.ca

- Canadian Cancer Society: the Canadian Cancer Society can provide information on cancer and cancer research http://www.cancer.ca
- National Institutes of Health: this website contains a database of clinical trials for many diseases and conditions http://www.clinicaltrials.gov

Clinical Trials Support Unit: the unit can provide a list of available trials at Princess Margaret.
Email ClinicalTrials-PMH@uhn.on.ca or phone 416-946-4501 extension 3835.

Living with an MPN

Finding out you have a rare and potentially life-threatening condition can be difficult and chronic diseases like MPNs can pose challenges to daily living

MPN symptoms

Fatigue

Fatigue is one of the most reported symptoms by patients diagnosed with an MPN. It can be difficult to get out of bed and to go about your daily routine. Currently there is no treatment for it but there are some tips you can work into your day to help manage your fatigue.

> Tracking your symptoms is useful to see how they change and whether your treatment is helping

Exercise – believe it or not, light to moderate exercise can improve fatigue[1]. Try to be active a little bit every day. It does not need to be very strenuous (especially if you have an enlarged spleen). Walking, yoga, swimming, tai chi or whatever is accessible to you and easy to do.

1. Scherber et al. (2016) Cancer 122(3):477-85

You should **talk to your doctor** first before starting any new fitness regime, build up your activity level gradually and rest when you need to. Your health care team can refer you to resources available at Princess Margaret or elsewhere to help improve fatigue.

Plan ahead – sometimes we plan too many activities to complete in one day. Do the most important things first, and spread it out during the week

Pace yourself – break up bigger tasks into smaller parts

Learn to say no – allow yourself to care for yourself

Learn to ask for help from others – it can help you accomplish more.

More resources are available at Princess Margaret to help you combat fatigue – see p.39 for more information.

Pruritis

Pruritis is a condition associated with MPN that causes intense itching (for some patients it happens after a shower). Creams with low pH may help, such as moisturizers, emollients, and barrier repair creams. A topical corticosteroid may help as well.

Satiety

Early satiety is feeling full after eating very little. This can happen if you have an enlarged spleen. One way to keep up your nutritional intake is to eat smaller meals throughout the day. A consultation with a dietician/nutritionist can be arranged during one of your visits.

Staying well

Taking care of your health will help reduce your chances of suffering a complication of your MPN (like a clot), improve your energy and help you feel the best you can.

Top tips for staying well

- Get or stay active
- Eat a healthy diet
- Drink plenty of water
- Manage stress
- Stop smoking
- Get to a healthy weight
- Moderate your alcohol intake

Support groups for MPN patients (p. 35) provide an opportunity to talk with others living with an MPN diagnosis.

Your state of mind

Mindfulness is taking the opportunity to reflect on all aspects of your life on a daily basis. It improves your mental well-being as well. Your mental well-being is just as important as your physical health. People with MPNs experience many stresses and feelings of anxiety, depression or grief are normal. These feelings can also change over time.

You may find it useful to talk with a friend, write your feelings down or talk to a psychologist (ask your doctor for a referral).

Support groups

When you've been diagnosed with a rare disease such as MF, PV or ET it's not unusual to feel isolated. But there are thousands of others who have walked the path before you who are willing to share their experiences and advice.

Below is a list of in-person MPN support groups operating Canada-wide and in Ontario. Groups in other provinces can be found on http://www.mpncanada.com

Support Groups

Canada-wide

Canadian MPN Network
Patient Advocacy Group
1-844-644-6766 (MPNN)

Cheryl Petruk
canadianmpnnetwork@gmail.com
780-940-6569

Ontario

Toronto
Contact: Kevin Brown
kb@iconhomes.com

Hamilton
Contact: John Clark
jwclark@gmail.com

Kitchener/Waterloo
Contact: Joanne McKinney
JMcki99425@aol.com

Windsor
Contact: Jane Fleming
Janefleming62@gmail.com

Ottawa
Contact: Phil Arner
phil.arner@rogers.com

Your Care at Princess Margaret

You may be referred to the Elizabeth and Tony Comper MPN Program at the Princess Margaret Cancer Centre for further investigation or to explore other treatment options for your MPN diagnosis.

Your first visit

Your initial appointment will last 2-4 hours.

At your first appointment you will have extensive blood work done, a detailed history will be taken and a physical examination will be done. You will have the opportunity to discuss your concerns, meet with the clinical trials team and a bone marrow biopsy may be done to rule out other hematological issues which may be contributing to your symptoms. You will also meet with the tissue bank coordinator to discuss the option of providing a sample of blood for future research.

After your first visit

After your appointment the MPN team will make a plan of care, which may involve your referring physician or family physician. This means that **you may receive some of your care at another centre or from your family physician.** This is our Shared Care philosophy and allows everyone to share in your care.

How often your visits are will depend on your treatment and symptom management. Your appointments could be as often as twice a month or as little as every 12 months.

Your MPN healthcare team

Your MPN team is comprised of staff physicians, fellows, a clinical nurse specialist, specialized oncology nurses and social workers. You will meet many of these team members during your clinic visits.

Contacting the MPN team

In an emergency: please go to your local Emergency Department.

On weekends and after hours: please call the Princess Margaret

main switchboard at: 416-946-2000 and ask for the nursing supervisor or leukemia doctor on call, or call your family physician.

Triage line: to contact the MPN team use the triage line to speak to a specialized oncology nurse regarding supportive counselling, symptom management, general concerns.
Dial: 416-946-2223 then select option 2.

Your clinical nurse specialist – for questions or concerns between appointments, including medication or treamtnet questions call Nancy Siddiq: 416-946-4501 ext. 3047

Your clinical trials nurse – if you participate in a clinical trial you will be given contact details.

Please have your MRN (medical records number) to hand when contacting your healthcare team.

Resources for MPN Patients

Resources at Princess Margaret

AYA

The Adolescent and Young Adult (AYA) program provides supportive counselling for the unique needs for patients 39 years of age and under. Your health care provider can make a referral at any time, or patients can refer themselves by email: aya@uhn.ca

Smoking Cessation

If you are a new patient and thinking about quitting, there is an electronic survey which can provide education about treatment options. Also ask your health care provider for the *"For The Smokers Who Want To Quit"* resource booklet. This booklet is available in clinics or at the Patient & Family Library at Princess Margaret.

Fatigue

Pamphlets

- Use Your Energy Wisely
- Helpful Hints for Better Sleep
- Relaxation Techniques for Stress Relief

To find these resources:

- Visit the Princess Margaret, Patient & Family Library (main floor)
- Go to www.theprincessmargaret.ca click on "Patients & Families", then click on "Patient & Family Library" to search for the resources.

Classes at Princess Margaret

- Relaxation Therapy Group Sessions
- Reclaim Your Energy: Coping with Cancer-Related Fatigue

  Attend this class in your language. You can book a medical interpreter for this class at no cost to you. To book, please call 416-581-8604. Five weekdays advance notice is needed.

For more information including times and location:

- Pick up the *'Patient Education & Survivorship Calendar of Events"* from the Princess Margaret Patient & Family Library.
- Go to www.theprincessmargaret.ca – click on "See Calendar" under "Classes & Activities" and search for the classes.

Patient & Family Library

The Patient & Family Library offers computer stations, books, brochures, DVDs, audio books, electronic books and CDs with reliable information about cancer, cancer treatment and care, support organizations and services and much more.

Trained staff and volunteers can help with your questions and help you to find information and resources. Our library staff and volunteers are friendly, professional and respectful.

Cancer information is available in several languages. The library has information available in English, Portuguese, Chinese, Italian and French. You can also request information in other languages.

Resources offered include:

- eBooks: Now you can read health, wellness and cancer survivorship books online! E-books can be accessed from your home computer, laptop, or mobile device. Access is free for UHN patients and families.
- Computer Access: There are computers with internet access for you to use in the Patient & Family Library. You can use these computers to check your email, stay in touch with friends and family, or search for health information.
- Tailored Information Packages: If you can't find what you are looking for in the library collection, you can request health information that is specific to your needs from the Patient & Family Library. Fill out a search form available online and bring it to the library or phone 416-946-4501 ext. 5383. The librarian or a trained volunteer will search for information for you. You can choose to have your information package sent to you by post or by email, or you can pick it up in person at the Patient & Family Library.

How to borrow material

You can borrow library materials for free for three weeks. You need a piece of identification such as your blue UHN card or a driver's licence to take material out of the library. You must be a patient, family member, friend or caregiver to borrow materials from the library.

Location

Main floor, across the atrium from the Murray Street entrance. There are also many satellite libraries in different clinics and centres throughout the hospital.

Hours and Contact

Monday – Thursday, 8:30 am – 4:30 pm;
Friday 8:30 am – 4:00 pm

Phone: 416-946-4501 ext. 5383

Email: patienteducation@uhn.on.ca

Other Resources

Canada's Food Guide

This website provides the most up to date information on choosing the healthy food options to improve your intake of essential vitamins and nutrients.
www.hc-sc.gc.ca/fn-an/food-guide-aliment/index-eng.php

Canada's Physical Activity Guide

This website has a variety of options of exercises which can help to improve mood and fatigue, and maintain healthy weight.
http://www.phac-aspc.gc.ca/hp-ps/hl-mvs/pa-ap/index-eng.php

Cancer Related Fatigue Video

This 10 minute video explores cancer related fatigue, and how it differs from normal fatigue, and some suggestions on how to improve fatigue.
https://www.youtube.com/watch?v=YTFPMYGe86s

For More Information

Want to learn more about your diagnosis? Looking for advice on living with an MPN? These organizations are excellent resources for MPN patients looking for more information.

Canadian MPN Group

The Canadian MPN Group is a charitable organization and a collaborative effort to improve care and research for patients with MPNs. They provide information about MPNs, clinical trials in Canada and links to support groups and other resources (http://mpncanada.com)

MPN Research Foundation

The MPN Research Foundation is a non-profit organization whose mission is to promote, fund and support the most innovative and effective research into the causes, treatments, and potentially the cure for polycythemia vera, essential thrombocythemia and myelofibrosis

MPD Voice

Based in the UK and established by a group of MPN patients, MPD Voice has an active online community and up-to-date information on essential thrombocythemia, polycythemia vera and myelofibrosis.

MPN Advocacy and Education International

MPN Advocacy and Education International is dedicated to providing information, support and resources for patients living with MPNs. The group conducts educational symposia, hosts webcasts, and direction to additional resources.

MPN Education Foundation

A U.S.-based non-profit organization run by volunteer MPN patients, the MPN Education Foundation hosts patient-doctor conferences to improve patient knowledge about their MPNs and to give patients the opportunity to meet with top MPN specialists.

MyelofibrosisAwareness.org (The MPN Coalition)

The MPN Coalition provides a forum for discussion of and action on the needs and challenges faced by those living with and affected by MPNs, including myelofibrosis. The Coalition aims to create greater awareness of these rare diseases and to enhance education and access to care.

Patient Power

Patient Power® is devoted to helping cancer patients and their families through knowledge. It is built on a library of programs organized into health centres, including an MPN health centre that provides helpful videos, news and tips

Canadian Cancer Society

The Canadian Cancer Society website is a good general resource on MPN, providing an overview of signs and symptoms, diagnosis and treatment options for polycythemia vera, essential thrombocythemia and idiopathic myelofibrosis.

The Leukemia & Lymphoma Society

The Leukemia & Lymphoma Society (LLS) is the world's largest non-profit health agency dedicated to helping patients with blood cancer and their families. LLS funds blood cancer research around the world and provides free information and support services to patients and their families.

National Organization for Rare Disorders (NORD)

For more than 30 years, NORD has led the way in voicing the needs of the rare disease community, driving supportive policies, advancing medical research and providing patient and family services for those who need them most.

MPN Research at Princess Margaret

Participating in Research

Though they are important, MPN research isn't only about clinical trials. There are many ways you can participate in MPN research at Princess Margaret.

Participating in any kind of research is always voluntary and will never affect your care.

Tissue Banking

Tissue banking involves giving a sample of tissue (this can be blood, nail clippings, bone marrow or hair), which is then preserved and stored for future medical research.

During your first visit to the MPN clinic you will be approached by our Tissue Bank Coordinator who will give you more information on this and ask if you want to participate.

Participating in Clinical trials

Clinical trials are important to develop new, safe and effective therapies for treating MPN. There are many types of clinical trials at Princess Margaret – some involve drugs or other therapies and some are observational only.

Your physician may recommend that a particular clinical trial may be suitable for you. The decision whether to participate should be carefully considered.

Every clinical trial has strict eligibility criteria that determine which patients can participate in the trial. There are also some factors that may exclude you from a particular clinical trial.

You can find more information on clinical trials on p.29

Frequently Asked Questions

What causes polycythemia vera (PV)?

The cause of PV is not fully understood. Almost everyone with PV has an acquired mutation in the JAK2 gene, however other unrecognized mutations may exist. The mutation occurs in a bone marrow stem cell, causing uncontrolled production of blood cells, especially red cells.

What causes myelofibrosis (MF)?

The cause of MF is unknown (idiopathic). Approximately 50% of patients with primary myelofibrosis (PMF) have a mutation in JAK2, 25% a mutation in the calreticulin (CALR) gene, and 5-10% a mutation in the MPL gene. However, the exact mechanism how mutations in JAK2 or MPL genes cause PMF is unknown.

What causes essential thrombocythemia (ET)?

The exact cause of ET is not known. Similar to primary myelofibrosis, mutations in the JAK2 gene are identified in 50% of patients, calreticulin mutations in 25% of cases and mutation of MPL in 1% of patients with ET.

Does a diagnosis of MPN mean a shorter lifespan?

With proper treatment and precautions, people with PV or essential ET are likely to have a normal lifespan. With MF, life expectancy differs with each case and largely depends on the overall health of the patient, how aggressively the disease behaves and how early treatment begins. The survival time ranges widely from 12 months in high-risk disease to more than 15 years in low-risk disease.

I was recently diagnosed with CALR mutation. What does it mean for me?

Mutation in calreticulin (CALR) is the second most common genetic defect (JAK2 is the first) in ET and MF. It is not commonly found in PV. CALR mutation is found more frequently in younger patients and is associated with higher platelet count and hemoglobin level. CALR patients with ET have blood clots less frequently; patients with MF tend to live longer compared with those with a JAK2 mutation.

If I have an MPN, are my children and siblings more likely to get it?

Most MPN cases are random — the changes in DNA that drive the development of MPNs are acquired during a person's lifetime. This means they are not inherited from parents and cannot be passed on to children. However, researchers have found that there is a predisposition towards developing MPN in some families, known as 'familial MPN'. Individuals in these families have a greater chance of developing MPN than normal[1].

What can I do to manage MPN in my daily life?

Maintaining a healthy lifestyle is an important first step.

- Get moderate regular exercise, such as walking or swimming, which will decrease your risk of blood clots and improve blood circulation.
- Avoid tobacco and alcohol.
- Keep your skin healthy and use lotion to keep it moist.
- Avoid hot tubs, heated whirlpools, hot showers and baths.
- Avoid scratching or damaging your skin, as this can increase the risk of infection.
- Avoid extreme temperatures. In hot weather, protect yourself from direct sunlight and remain well hydrated. In cold weather, wear warm clothing, particularly on your hands and feet.
- Inspect your legs regularly and watch for leg sores. Patients with MPN, and in particular PV, have impaired blood circulation.

1. Landgren O, et al. Blood 2008;112:2199-2204

What physical activity is advised if I have an enlarged spleen?

Moderate exercise can safely raise your heart rate and improve blood flow to your body. But if you experience pain in the left part of your stomach because of an enlarged spleen, start your exercises at a lower intensity level.

Avoid situations where you may be at risk of injury, such as contact sports. If you are injured, particularly in the abdominal area, seek treatment right away and tell the person treating you that you have an MPN.

Should I make any changes to my diet?

In general, people with MPN should adopt the same healthy eating habits recommended for everyone. Adopt a lower fat diet with less red meat and less salt, in regular portions with a balance of fruits and vegetables. Use healthy oils like olive, sunflower and canola oil, and reduce animal fats. Stay well hydrated, in particular in warm weather. Avoid refined food, caffeine, alcohol and tobacco.

Should I take vitamin and mineral supplements or other food supplements?

There are no studies suggesting that any herb or nutritional supplement offers therapeutic benefits to people with MPN.

Patients with PV who undergo phlebotomies should avoid iron supplementation and high amounts of vitamin C. Because of increased risk of blood clots, most hematologists recommend against estrogen-based hormonal contraception or hormone replacement.

Should I see a physiotherapist or naturopathic doctor?

There are no special recommendations about physiotherapy or naturopathic therapy. If you decide to consult a physiotherapist or naturopath, look for a person with experience in caring for people with chronic illness.

Is it safe for me to fly?

Long-distance travel is considered a risk factor for blood clots forming in veins. However, while people with MPNs are prone to blood clotting, travel by air is not prohibited.

Avoid alcohol and sleeping pills and do leg exercises during the flight. You may need graduated compression stockings, pre-flight aspirin or blood thinners. Discuss these options with your hematologist.

Is it safe for me to get vaccinations, like flu and zoster?

If you have had your spleen removed, you must get vaccinations to prevent future infections. You should avoid live vaccines, but should get the annual flu vaccine. Ask your hematologist for advice.

How does phlebotomy work?

Phlebotomy, also known as bloodletting or venesection, is the mainstay of PV treatment. It is used to remove excess iron and maintain low body iron stores. Iron is used by bone marrow stem cells to produce new red blood cells. By reducing the amount of available iron, called iron deficiency, phlebotomy controls excessive red blood cell production.

How can I get in touch with a patient support group?

There are many support groups for people with MPN. You can find support groups in Canada on p.35.

Glossary

Abdomen

The area between the end of the rib cage and the beginning of the pelvic bone (hip). The major organs in this area are liver, spleen, stomach, pancreas, gallbladder, small intestine, and colon.

Abdominal pain

Pain that is felt in the area of the abdomen.

Acute myeloid leukemia

A quickly progressive malignant disease in which there are too many immature blood forming cells (red blood cells, white blood cells, platelets) in the blood and bone marrow.

Allogeneic bone marrow transplant

A procedure in which new healthy stem cells are transferred from a donor's bone marrow, or from a donor's peripheral blood, to the recipient. Donors can be related or not related to the recipient (another name for hematopoietic stem cell transplant (HSCT)).

Anagrelide

A drug that helps to reduce platelet count only. It is only recommended if hydroxyurea therapy fails. Anagrelide does not help with splenomegaly. It is not recommended for patients with heart conditions.

Anemia

A medical condition in which the red blood cell count (hemoglobin) is lower than normal.

Aspirin therapy

Low dose aspirin is used keep the blood thin to prevent blood clots forming.

Blood clot

Forms when the blood cells (red blood cells, white blood cells, or platelets) start to stick together and form a clot (thrombus). A blood clot becomes dangerous if it travels through the blood

stream and blocks blood flow to an area of the body, or to an organ (such as the brain, heart, liver, lungs or kidneys).

Blood transfusion

The transfer of blood or blood products (red blood cells, platelets, plasma) from a donor to a recipient intravenously. Transfusion is used to replace blood loss due to accidents or low blood counts due to illness.

Bone marrow

The spongy tissue found inside the long bones (femur in the thigh), sternum (middle of rib cage), or pelvis (hip). Hematopoietic stem cells are formed in the marrow that produce red blood cells, white blood cells (infection fighting cells), and platelets.

Busulphan

Busulphan is a chemotherapy drug which can reduce high red blood cell count in polycythemia vera (PV) patients. Busulphan is only used as a last line of therapy due to its potential to cause leukemia.

Calreticulin (CALR)

This protein has many responsibilities to ensure proteins can function normally in different cells (e.g. muscle cells, nerve cells, brain cells etc). This gene is known to be mutated in some patients with essential thrombocythemia (ET), and primary myelofibrosis (PMF), but it is not yet well understood.

Chemotherapy

Therapy for cancer using chemicals that stop the growth of cancer cells.

Chronic

A disease or illness where symptoms continue for a long period of time. Myeloproliferative Neoplasms (MPNs) are considered chronic conditions.

Complementary therapy

Alternative therapies such as acupuncture, yoga, homeopathy and naturopathy, which can be used alongside conventional therapies to provide symptom control.

Complete Blood Cell count (CBC)

This test measures the level for hemoglobin (red blood cells), leukocytes (white blood cells), platelets and a several other important values that help to determine a person's health status. These can also be abbreviated as: Hb (hemoglobin), WBC (white blood cells) and plts (platelets).

Cytokine

Cytokines are proteins that help to regulate the immune system, stem cell production (hematopoeisis) and inflammation.

Cytoreductive therapy

Therapy to reduce the number of malignant cells.

Early satiety

Feeling full after eating a small amount of food.

Erythrocytosis

Increased number of red blood cells (hemoglobin).

Erythropoietin

A hormone produced by the kidneys that promotes the production of red blood cells in the bone marrow.

Essential thrombocythemia

A rarely-acquired MPN characterized by an overproduction of platelets, which can lead to increased clots (thrombosis) and increased bleeding (hemorrhage).

Fatigue

A lack of energy and motivation. Fatigue can be acute and come on suddenly, or be chronic and persistent.

Fever

Increase in body temperature over 38°C / 100.4°F.

Heart attack

Damage to or death of the heart muscle due to loss of blood supply to the coronary artery.

Hematocrit (Hct)

The percentage of red blood cells in the total blood volume.

Hematologist

A doctor specially trained in hematology.

Hematology

The diagnosis, treatment and prevention of diseases of the blood and bone marrow.

Hematopoeisis

The process of forming new blood cells. All blood cells begin as hematopoietic stem cells in the bone marrow before developing into more specialized types of blood cell (red blood cells, white blood cells or platelets).

Hematopoeitic stem cells (HSCs)

The stem cells in the bone marrow that form all new blood cells (red blood cells, white blood cells and platelets).

Hematopoietic stem cell transplant (HSCT)

A procedure in which healthy hematopoietic stem cells are transferred from a donor's bone marrow, or from a donor's peripheral blood, to the recipient (patient). Donors can be related or not related to the recipient (another name for allogeneic stem cell transplant or allogeneic bone marrow transplant).

Hepatomegaly

An enlarged liver.

Hydroxyurea

An antimetabolite (interferes with cell reproduction) that can reduce blood counts.

Hyperviscosity

Thickening of the blood.

Interferon

A cytokine (see cytokine) released by the immune system when the body has an infection or cancer cells are invading.

Interferon can be given by injection to help stop the cancer cells from growing and to signal other immune cells to attack cancer cells.

Janus Kinase (JAK2)

JAK2 is a protein that exists in all people. It forms a communications pathway for messages travelling inside the cell. JAK2 has a role in the production of normal healthy blood cells.

Liver

An organ in the upper right abdomen that aids in digestion and removes waste products and worn-out cells from the blood.

Marrow fibrosis

When the spongy tissue in the marrow forms scar tissue and is no longer able to produce hematopoeitic stem cells.

MPL

A protein that is mutated in some MPN patients. It is involved in the production of platelets.

Myelofibrosis (MF)

Fibrosis or spontaneous scarring of the bone marrow. It is characterized by significant anemia (low red blood cell count / low hemoglobin) and an enlarged spleen.

Myeloproliferative neoplasms (MPNs)

Myeloproliferative neoplasms are diseases of the blood and bone marrow and are sometimes referred to as blood cancers. Four main types make up around 95% of MPNs: primary myelofibrosis (MF), essential thrombocythemia (ET), polycythemia vera (PV) and chronic myeloid leukemia (CML).

Neutropenia

A condition in which the number of neutrophils (a type of white blood cell) in the bloodstream is decreased

Night sweats

Severe hot flushes that occur at night and result in a drenching sweat.

Phlebotomy

A procedure where a set amount of blood is removed to decrease elevated haemoglobin (red blood cells).

Platelet

An irregular, disc-shaped cell in the blood that assists in blood clotting.

Platelet count

The calculated number of platelets in a volume of blood. The normal range is 150-450 x 10^9 per litre (150-450 billion per litre).

Polycythemia vera (PV)

A myeloproliferative neoplasm that characterised by an over-production of red blood cells.

Post-ET myelofibrosis

Transformation of essential thrombocythemia to myelofibrosis.

Post-PV myelofibrosis

Transformation from polycythemia vera to myelofibrosis.

Pruritus

Itching that can be caused by MF.

Radiotherapy

High energy rays are used to damage cancer cells and stop them from growing and dividing.

Red blood cell (erythrocyte)

The blood cell that carries oxygen.

Ruxolitinib (Jakavi®)

A commercial drug that helps to decrease spleen size and symptoms associated with PV and MF. Also known as a JAK2 inhibitor.

Spleen

An organ located in the upper left part of the abdomen near the stomach (usually the size of a fist). It acts as a reservoir for

blood and filters old blood cells. In MPNs the spleen may try to assist in hematopoeisis (blood cell production) when the bone marrow is unable to.

Splenectomy

Surgical removal of the spleen.

Splenomegaly

An increased spleen size.

Stem cells

Stem cells are cells that have the potential to develop into many different specialized cell types.

Stroke

Lack of blood flow to the brain due to a blockage, resulting in brain cell damage or death.

Thrombocythemia (thrombosis)

An abnormally high number of platelets in the blood. Greater than 450 x 10^9 per litre (450 billion per litre).

Thrombocytopenia

A lower than normal number of platelets in the blood. Lower than 150 x 10^9 per litre (150 billion per litre).

Thrombosis

The formation or presence of a blood clot in a blood vessel.

White blood cell (leukocyte)

One of the cells the body makes to help fight infections. The two most common are lymphocytes and neutrophils

Yoga

A way of life that includes dietary modifications, physical exercise and meditation

Notes

Notes

Feedback

I liked….

What could be better…

Anything else…

The development of this resource is supported
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Help us improve!

Your feedback helps improve this booklet for future patients

A feedback form is at the end of this booklet, or you can talk to any of our MPN staff or email: nancy.siddiq@uhn.ca

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