Myeloproliferative neoplasms

Your guide to best cancer care

About this guide

Being told you have or could have a myeloproliferative neoplasm (MPN) can be overwhelming. A lot can happen quickly, and you might have many questions. There are three main types of MPNs which include essential thrombocythaemia (ET), polycythaemia vera (PV), and myelofibrosis (MF). This resource can help to guide you, your family, and friends through this experience.

Information and support

For more information about MPN, visit the Leukaemia Foundation website **https://www.leukaemia.org.au/blood-cancer/myeloproliferative-neoplasms/**.

Leukaemia Foundation: To speak with an experienced healthcare professional who can provide you with disease-specific information, answer your questions, talk through your concerns, and connect you to blood cancer support groups, call **1800 620 420** or visit **www.leukaemia.org.au**. The team can also help with practical concerns such as accommodation close to treatment, transport to appointments, and financial assistance.

MPN Alliance Australia: is a small team of volunteer Australian MPN patients who provide evidence-based information and support for MPN patients via their website **www.mpnallianceaustralia.org.au**. The website includes an information and support section for newly diagnosed patients, the latest MPN articles, treatment information, and several support options, including from other MPN patients.

Cancer Council: for information and support, call **13 11 20** to talk to an experienced healthcare professional or visit **www.cancer.org.au**.

More information is available in the 'Resources' section of the optimal care pathway for myeloproliferative neoplasms. **www.cancer.org.au/OCP**.

Translating and Interpreting Service (TIS): If you need a translator, call TIS on **13 14 50** or visit **www.tisnational.gov.au**.

Initial tests and referrals

Symptoms

Many patients with an MPN may be unaware of having symptoms, and your condition may be first noticed by your doctor as unusual results in a routine blood test. If you have presented to your general practitioner (GP) or other primary care physician with symptoms, they will do a check-up to see if they can find out what is making you unwell. They will ask you about any symptoms you might be having such as fatigue, concentration problems, fevers or night sweats, abdominal discomfort, weight loss, bone pain, itchy skin (particularly after exposure to water), gout, or burning pain in the hands and feet in response to heat. Please discuss any symptoms with your doctor.

Initial tests you may have:

Physical exam. Your doctor will check to see the cause of symptoms such as those noted above or unusual blood results.

Blood tests. A sample of your blood is collected and sent for a full blood examination and potentially other tests to see if there are markers for MPN present in the blood.

Referrals

If your GP/physician thinks you may have an MPN, they'll refer you to a haematologist or medical oncologist (specialist) at a public hospital or in private practice for more tests. Haematologists are highly trained in conditions that affect blood and bone marrow.

It can help to have a family member, carer, or friend attend appointments with you.

Let your doctor know what is most important to you in a specialist (e.g., that they specialise in MPN, cost, location, bedside manner, expertise). However, we recommend prioritising MPN expertise over other criteria to ensure best care and outcomes.

Timeframes

The timing of your specialist appointment will depend on your initial results of tests performed by your GP/physician. For a small number of patients, particularly those with a very high haematocrit (large volume of red blood cells), an urgent appointment, venesection (drawing a controlled amount of blood from the bloodstream) or hospital admission may be required. But for most people with no urgency, an appointment **within eight weeks** is appropriate.

If you can't get an appointment within this timeframe, follow up with your referring GP/physician.

Ask your GP to recommend trusted sources of information and support — you can share these with your family and friends too.

Talk to your GP/specialist about how to manage any other health conditions you may have.

Questions you might want to ask your referring GP/physician

- Are there any symptoms that I should watch out for while I am waiting for my appointment to see a specialist?
- Can I choose the specialist I see?
- Can I choose whether I see a public or private doctor?
- What are the differences of being treated in the public versus private health system?
- What emotional and mental health support services are available and how do I access them?

If you are concerned about costs in the private health system, it is recommended that you ask for a referral to a public hospital, especially if you have PV and may need regular venesections or if you need transfusions and bone marrow biopsies which can carry out of pocket costs even with private health insurance.

It is very helpful to request and keep copies of your blood test results, as well as reports of any scans or biopsies, etc. You are then able to take them with you when you see any other health provider for other health conditions, so that your care may be best suited to your particular needs.

It's a good idea to keep a written diary or digital record of treatment details and appointments with your GP and specialists. You can ask permission to record appointments. It's also helpful to bring a list of questions and ask for a written treatment plan.

Don't ignore new signs and symptoms. Alert your GP or specialist. Trust yourself. It's OKAY to be persistent.

Travel to access tests, treatment, and care

You may regularly have to travel to see a specialist, undergo tests, or access treatment, especially if you live in a rural or regional area. In some cases, you may have to leave home for an extended period of time. Most MPN patients require ongoing treatment and management via regular blood tests and visits to their haematologist.

Mental health and emotional wellbeing

Even though most MPNs are a chronic blood cancer, a blood cancer diagnosis can affect your mental and emotional wellbeing. Patient support organisations, or another health professional such as a psychologist or social worker can help you develop strategies to cope. They can help you access a mental health treatment plan if required. You can ask your haematologist about whether there are any support groups available in your local area or online.

Diagnosis and treatment planning

The haematologist/specialist will do more tests to determine if you have an MPN to help determine the best treatment for you.

You may have one or a combination of these tests:

- **Clinical assessment and screening** to check for cardiovascular risk factors, for example.
- **Blood tests.** A blood sample is taken for a full blood evaluation and blood film examination. Other blood tests include tests for kidney and liver function, and for mutations relating to MPNs.
- **Imaging.** Chest X-ray and abdominal ultrasound may be requested.
- **Bone marrow testing.** A bone marrow sample is collected from the hip using a thin needle. You may have a bone marrow aspirate and/or a bone marrow biopsy. The fluid portion of the bone marrow will be sampled for a bone marrow aspirate and the solid portion will be sampled for a bone marrow biopsy. Samples are then sent to a laboratory for examination.

Timeframes

You should have most results back **within eight weeks**.

Initial questions you might want to ask

- What are MPNs and where can I find more information about them?
- What tests will I need and why? Do I need a bone marrow biopsy?
- Will my follow-up appointments be face-to-face or by tele/video health or a combination? How frequent are they?
- How much will appointments and tests cost and how much will I have to pay myself?
- What type of MPN do I have?
- What are my chances that the MPN (if ET or PV) can become myelofibrosis (MF)?
- What are my chances that the MPN can become leukaemia?
- What are the symptoms and signs that the MPN may be changing?
- What is my risk of blood clots, what can I do to minimise it and how would I recognise it?
- Can I choose where I have treatment, and what are my treatment options?
- Will I need to start treatment straight away?
- How long will I need to have treatment?
- How will my disease be monitored?
- Is there information that I can share with my family or friends?

Treatment

Your specialist will talk to you about your treatment options for your MPN. You may need more than one treatment type to get the best results, and you will require monitoring and are likely to require ongoing treatment to manage your MPN for the rest of your life. Some patients will initially be managed with a ‘Watch and Wait’ approach which involves regular blood testing and appointments with your haematologist to check for symptoms or changes in your blood test results.

You may also have one or a combination of these treatments:

- **Anti-platelet & anti-coagulant therapy.** Small daily doses of antiplatelet medication like aspirin or an anticoagulant medication like warfarin for most people with MPNs prevent platelets from clumping together and reduce the risk of blood clots.

- **Venesection/phlebotomy.** Used for the treatment of PV and is a procedure where a controlled amount of blood is removed from the bloodstream to reduce the high red blood cell count.
- **Transfusions.** If you have low red blood cell counts, you may need a blood transfusion which is a slow injection of red blood cells into a vein (intravenously or IV). Similarly, if you have a low platelet count, you may need a platelet transfusion where you will be given a slow injection of platelets into a vein.
- **Cytoreductive therapy.** Considered for high-risk patients with MPN. These medications include chemotherapy drugs, immunotherapy drugs such as Interferon, platelet reducing agents such as Agrylin, and JAK inhibitors such as Ruxolitinib, which help to control the number of red and white blood cells and platelets in the bloodstream as well as the size of the spleen.
- **Allogeneic stem cell transplant.** This is reserved for high-risk patients only; your haematologist will discuss if this is an option for you.

For more information visit **www.cancer.org.au/cancer-information/treatment**.

Supportive care (treatment or services that support you throughout your cancer experience) are also available, often delivered through a multidisciplinary team that may include nurses, pharmacists, psychologists, physiotherapists, and dietitians. Don't hesitate to ask your GP or haematologist for contact details, especially for cancer psychologists.

Timeframes

By evaluating your symptoms and test results, your specialist will determine when your treatment should start. For most people this will generally be **within eight weeks** of the decision being made. For people who are at a high-risk for blood clots, treatment should **begin immediately**, usually starting with taking anti-platelet or anti-coagulant medications and venesections if haematocrit is very high in PV patients. There may be instances where your treatment could be delayed beyond this timeframe. If this occurs your specialist will discuss this with you.

Clinical trials

Your specialist may recommend participating in a clinical trial (or you can ask if you are eligible for any clinical trials). Clinical trials often provide access to promising new treatments that are not yet available to the general public. Many people with cancer are now living longer, with a better quality of life, because of clinical trials.

For more information about the types of trials available and information on participating in a trial, visit **www.australiancancertrials.gov.au** or search the Australian New Zealand Clinical Trials Register **www.anzctr.org.au**.

Complementary therapies and other medications

It is important that you speak to your healthcare team about any dietary supplements, vitamins, herbal remedies, and other medications that you use or would like to use, including prescription and over-the-counter medicines (e.g., paracetamol or ibuprofen). Some medicines, complementary therapies, vitamins, and foods might not work well with your treatment.

Fertility and sexual health

Blood cancer and blood cancer treatment may cause fertility problems for both men and women. This will depend on age, the type of blood cancer, and the treatment received. If this is relevant to you, get advice from your treating team about contraception and having children whilst undergoing treatment. If you are approaching or experiencing menopause, whether you are able to take hormone replacement therapy is an important discussion to have with your haematologist. Diagnosis and treatment may affect your sex life or overall sexual health. For support and, if necessary, referral to counselling services, contact the Leukaemia Foundation on **1800 620 420** or visit **www.leukaemia.org.au** and type 'relationships and sex' in the search bar.

You can ask for more time to think about your treatment options.

You can ask your GP for a referral to another specialist for a second opinion.

Questions you might want to ask

- What are the treatment options available to me and what is their impact on MPN progression?
- What are my options if the MPN progresses?
- What will happen if I don't have treatment?
- Will I need to be on treatment, have regular blood tests, and be monitored for the rest of my life?
- What will treatment cost and how much of the cost will I have to pay myself?
- Who is in my treatment team and who is the main contact person if I have any questions or feel unwell between appointments?
- What side effects could I have from treatment and how should I manage them?
- How might the recommended treatment interact with other medications I am already taking?
- What if I need to have surgery or other health procedures? Do I need to stop my treatment?
- What do I need to know if I am travelling and on long flights to manage the risk of blood clots?
- What activities/exercise will help me to live well with an MPN?
- Is there any specific diet I should follow or foods or drink to avoid now that I have an MPN?
- How will my MPN affect my day-to-day life, and can I still work?
- Will treatment affect my ability to have children, and if yes, what options should I consider?
- Should I get a medic alert?

Decisions about cost

You may have to pay for some appointments, tests, medications, accommodation, travel, or parking.

Speak with your GP, specialist, or private health insurer (if you have one) to understand what is covered and what your out-of-pocket costs may be. This is important as your treatment will be ongoing, for example if you need regular venesections or blood transfusions.

If you have concerns about costs, talk to your healthcare team or a social worker about:

- being bulk-billed or being treated in the public system
- help with accommodation and/or transport
- the possible financial impact of your MPN and financial support schemes you may be able to access
- how your MPN might impact your ability to work.

For more information about financial and practical support call the Cancer Council on **13 11 20** or visit **www.cancer.org.au/support-and-services/practical-and-financial-assistance**, or contact the Leukaemia Foundation on **1800 620 420** or visit **www.leukaemia.org.au/how-we-can-help/practical-support-services/**.

Ongoing Care

A priority for treating MPNs is optimising symptom management and quality of life for patients through using combined medical approaches and supportive therapies to support your physical, psychological, nutritional, and general wellbeing. Comprehensive support and side effect management should be part of your routine ongoing care. It is important to ask your GP and follow health advice on vaccinations including for COVID-19 and other infections such as influenza or shingles.

Care management plan

You will need regular ongoing follow-up appointments and care, so your specialist and healthcare team will work with you to make a chronic disease management plan for you and your GP. This may need to be updated over time if your needs change. This plan will explain:

- who your main contact person is on a day-to-day basis
- how often you should have check-ups and what tests this will include
- understanding and dealing with the symptoms of the disease and the potential side effects of treatment
- how to get help quickly if you have an urgent problem, e.g., blood clots and unusual bleeding
- how to look after your overall health and wellbeing
- what healthcare and community support services are available to you and how to access them.

Your specialist and healthcare team will talk to you about your needs and can refer you to other health professionals and community support services. Other information you get might be about:

- the symptoms of the disease and how to manage them
- the side effects of treatment and the specialists you may need to see
- how to make healthy lifestyle choices to give you the best chance of staying well.

For more information visit **www.cancer.org.au/cancer-information/after-a-diagnosis/after-cancer-treatment**.

Questions you might want to ask

- Who should I contact if I'm feeling unwell?
- What can I do to be as healthy as possible?
- Where can I get more help?

Living well with a chronic disease

Patients with MPN have an increased risk of other malignancies and therefore should have preventative strategies and appropriate screening (such as bowel, cervical, breast, and prostate cancer screening).

Monitoring of blood counts, renal, and liver function and tolerance to treatment should be undertaken by practitioners who have experience in managing patients with MPNs. These can be tailored to the patient's treatment regimen and risk factor profile.

Other treatment options may include:

- standard anti-leukaemia strategies (considered on a case-by-case basis)
- clinical trials
- referral to a specialist bone marrow (stem cell) transplant centre
- supportive care and symptom control.

Questions you might want to ask

- What is my prognosis?
- Are there alternative treatment options?
- Can I be cured?
- Are there clinical trials available to me?
- What financial, practical, or emotional support is available?

Advance care planning

Your GP or healthcare team may talk with you, your family, and carer about future treatment and medical needs, if or when needed.

Advance care directive

Sometimes known as a living will, an advance care directive is a legally binding document that you prepare to let your family and medical team know about the treatment and care you might want or not want in case you become too unwell to make those decisions yourself. For more information visit **www.advancecareplanning.org.au**.

Symptom and palliative care management

Your specialist may refer you to palliative care services, but this doesn't always mean end-of-life care. Today people can be referred to these services much earlier if they're living with blood cancer or if their blood cancer returns or progresses. Palliative care can help you to live as well as you can including managing pain and symptoms, psychosocial support, and assistance in identifying care goals. This care may be at home, in a hospital, or at another location you choose. Speak to your GP or haematologist/specialist or visit **www.palliativecare.org.au**.

Making treatment decisions

You may decide not to have treatment at all, or to only have some treatment to reduce pain and discomfort. You can always change your mind about your treatment. Just talk to your specialist. For more information visit **www.cancer.org.au/cancer-information/treatment/advanced-cancer-treatment**.

Questions you might want to ask

- What can be done to reduce my symptoms?
- What extra support can I get if my family and friends care for me at home?
- Can you help me to talk to my family about what is happening?
- What support is available for my family or carer?
- Can I be referred to a community support service?
- What support services are available to me to help manage the ongoing nature of my blood cancer?

What can I do to help myself now that I have an MPN?

Many people are living longer with cancer, and many people with MPNs can have an almost normal life expectancy. It is important that people receive ongoing care and support in finding a 'new normal', managing general health, the effects of the cancer and/or treatment, and coping with the emotional impact of cancer and fear of progression. It is important to take good care of yourself and there are many things you can do to improve your general health and wellbeing including, eating a balanced diet, maintaining a normal weight, exercising, quitting smoking, and considering wellness activities such as yoga and meditation.

Helpful links

- Information for living well with an MPN: **www.mpnallianceaustralia.org.au**
- Leukaemia Foundation Emotional Support Services: **www.leukaemia.org.au/how-we-can-help/emotional-support-services/**
- Leukaemia Foundation Health and Wellbeing Services: **www.leukaemia.org.au/how-we-can-help/health-and-wellbeing-support-services/**
- Leukaemia Foundation Support Groups and Webinars: **www.leukaemia.org.au/get-involved/events/**.

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This resource is based on information from the optimal care pathway for people with myeloproliferative neoplasms (1st edition), available at **www.cancer.org.au/OCP**.