

TREATMENT UPDATE:

Myeloproliferative Neoplasms (MPNs)

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Treatment Update: Myeloproliferative Neoplasms (MPNs)

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Myeloproliferative neoplasms (MPNs) are a group of rare cancers originating in the bone marrow.

Bone marrow is the spongy tissue inside large bones. It manufactures stem cells, which in turn produce white blood cells that fight infection, red blood cells that carry oxygen and platelets that help with blood clotting. An MPN occurs when there is an imbalance in the production of these cells or platelets.

The primary types of MPNs are polycythemia vera (PV), myelofibrosis (MF) and essential thrombocythemia (ET).

Polycythemia vera (PV)

Polycythemia vera develops when bone marrow produces too many red blood cells, which can lead to a thickening of the blood. The vast majority of people with PV have a mutation (change) in the JAK2 gene that leads to the over-production of red blood cells.

As PV progresses, it can cause headaches, shortness of breath, bleeding, dizziness, itchiness and/or an enlarged spleen. PV can also increase the risk of the development of blood clots.

A primary goal of therapy for PV is to maintain a hematocrit level below 45%, which is proven to decrease the person's risk of forming a blood clot. The hematocrit level is the ratio of the volume of red blood cells to the total volume of blood. Phlebotomy (the use of a needle to take blood from a vein) is the therapy of choice to maintain hematocrit levels below 45%.

If phlebotomy alone does not achieve the desired hematocrit level, there are medications that can reduce the number of red blood cells in the bloodstream. These medications include hydroxyurea (Droxia, Hydrea), interferon alfa-2b (Intron A), ruxolitinib (Jakafi) and busulfan (Busulfex, Myleran).

The investigational drug rusfertide is currently being studied in clinical trials. According to research results, twice-weekly injections with rusfertide effectively controlled hematocrit levels and significantly reduced the need for phlebotomy.

Myelofibrosis (MF)

Myelofibrosis occurs when bone marrow cells called fibroblasts make too much fibrous (scar) tissue. As this scar tissue builds up, blood cells can no longer develop properly inside the bone marrow. This can cause anemia (a low number of red blood cells), a low platelet count, and a higher risk of developing infections. People with MF often have an enlarged spleen, which can cause pain or discomfort in the abdomen or below the ribs.



Some people with MF are asymptomatic (have no symptoms) and do not require treatment. Instead, their condition is closely monitored, which is called active surveillance. Treatment, if required, depends on the person's symptoms.

Medications

Medications prescribed for the treatment of MF are designed to reduce spleen enlargement and relieve symptoms.

- Ruxolitinib (Jakafi), fedratinib (Inrebic) and pacritinib (Vonjo) target mutations in the JAK2 gene and block its faulty processes.
- Momelotinib (Ojjaara), a treatment indicated for intermediate- or high-risk MF with anemia, was approved by the U.S. Food and Drug Administration (FDA) in September 2023. Momelotinib is a JAK1/JAK2 and ACVR1 inhibitor.
- Hydroxyurea (Droxia, Hydrea), epoetin alpha (Procrit) and darbepoetin alpha (Aranesp) are chemotherapies.

Splenectomy

The spleen helps fight infection by producing white blood cells. It also stores red blood cells and platelets. The surgical removal of the spleen (splenectomy) is considered for the treatment of symptomatic MF that is not responding to medication or radiation therapy administered to reduce spleen size.

Stem Cell Transplant

A stem cell transplant is a procedure in which diseased bone marrow is replaced with healthy bone marrow. In an allogeneic stem cell transplant, stem cells (blood cells that divide to make new blood cells within the bone marrow) are removed from a donor. Donors are often a close relative such as a brother or sister, but sometimes can be an unrelated volunteer who has a compatible bone marrow.

The person receiving the stem cell transplant is given high doses of chemotherapy to destroy abnormal cells in the bone marrow. The stem cells are then infused (transplanted) into the body, where they start working in the bone marrow to produce healthy blood cells.

Immunosuppressive drugs are typically given for a period of time after the transplant. This is to prevent the rejection of donor cells and to ensure the transplanted cells do not cause an autoimmune disease, such as graft-versus-host disease (GVHD). These drugs include cyclosporine and tacrolimus.

There is a high level of risk associated with an allogeneic stem cell transplant, and it is typically not the initial treatment approach for MF.

Essential thrombocythemia (ET)

Essential thrombocythemia occurs when the bone marrow produces too many platelets, which can cause blood clots that affect vital organs throughout the body.

People with ET who have no symptoms usually do not require treatment. Medication may be prescribed if the platelet count is very high, if the person has a history of blood clots, is older or has risk factors for heart disease. Platelet-lowering medications include hydroxyurea (Droxia, Hydrea), anagrelide (Agrylin) and interferon alfa-2b (Intron A).

In emergency situations where the platelet count must be reduced quickly, excess platelets can be removed from the bloodstream by a procedure called plateletpheresis. Blood is drawn from a needle placed in a vein and passed through a machine called a cell separator, which spins the blood and removes the excess platelets.

The Importance of Clinical Trials

Clinical trials are the standard by which we measure the safety and effectiveness of new treatments and the quality of life of patients as they receive those treatments. For this reason, doctors and researchers urge people with cancer to take part in clinical trials.

Your doctor can guide you in making a decision about whether a clinical trial is right for you. Here are a few things that you should know:

- Often, people who take part in clinical trials gain access to and benefit from new treatments.
- Before you participate in a clinical trial, you will be fully informed as to the risks and benefits of the trial, including any possible side effects.
- Many clinical trials are designed to test a new treatment against a standard treatment to find out whether the new treatment has any added benefit.
- Participation is voluntary and does not affect your access to treatment in other settings. You can stop taking part in a clinical trial at any time for any reason.

Treatment Side Effects

All cancer treatments can cause side effects. It's important that you report any side effects that you experience to your health care team so they can help you manage them. Report them right away—don't wait for your next appointment. Doing so will improve your quality of life and allow you to maintain your treatment plan. It's important to remember that not all patients experience all side effects, and patients may experience side effects not listed here.



The side effects of treatment depend on the type and dose of drugs given and the length of time they are used. They can include:

- Increased risk of infection (from having too few white blood cells)
- Easy bruising or bleeding (from having a low platelet count)
- Changes in memory or thinking
- Peripheral neuropathy (numbness or tingling in hands and feet)
- Weight gain
- Hair loss
- Headache
- Confusion
- Coordination difficulties

General Side Effects

Some side effects may occur across treatment approaches. This section provides tips and guidance on how to manage these side effects should they occur.

Managing Digestive Tract Symptoms

Nausea and vomiting

- Avoid food with strong odors, as well as overly sweet, greasy, fried or highly seasoned food.
- Eat meals that are chilled, which often makes food more easily tolerated.
- Nibble on dry crackers or toast. These bland foods are easy on the stomach.
- Having something in your stomach when you take medication may help ease nausea.

Diarrhea

- Drink plenty of water. Ask your doctor about using drinks such as Gatorade which provide electrolytes. Electrolytes are body salts that must stay in balance for cells to work properly.
- Over-the-counter medicines such as loperamide (Imodium A-D and others) and prescription drugs are available for diarrhea but should be used only if necessary. If the diarrhea is bad enough that you need medicine, contact a member of your health care team.
- Choose foods that contain soluble fiber, like beans, oat cereals and flaxseed, and high-pectin foods such as peaches, apples, oranges, bananas and apricots.
- Avoid foods high in refined sugar and those sweetened with sugar alcohols such as sorbitol and mannitol.

Loss of appetite

- Eating small meals throughout the day is an easy way to take in more protein and calories, which will help maintain your weight. Try to include protein in every meal.
- To keep from feeling full early, avoid liquids with meals or take only small sips (unless you need liquids to help swallow). Drink most of your liquids between meals.
- Keep high-calorie, high-protein snacks on hand such as hard-boiled eggs, peanut butter, cheese, ice cream, granola bars, liquid nutritional supplements, puddings, nuts, canned tuna or trail mix.
- If you are struggling to maintain your appetite, talk to your health care team about whether appetite-building medication could be right for you.

Managing Fatigue

Fatigue (extreme tiredness not helped by sleep) is one of the most common side effects of many cancer treatments. If you are taking a medication, your doctor may lower the dose of the drug, as long as it does not make the treatment less effective. If you are experiencing fatigue, talk to your doctor about whether taking a smaller dose is right for you.

There are a number of other tips for reducing fatigue:

- Take several short naps or breaks during the day.
- Take short walks or do some light exercise, if possible.
- Try easier or shorter versions of the activities you enjoy.
- Ask your family or friends to help you with tasks you find difficult or tiring.
- Save your energy for things you find most important.

There are also prescription medications that may help, such as modafinil. Your health care team can provide guidance on whether medication is the right approach for your individual circumstances. Fatigue can be a symptom of other illnesses, such as anemia, diabetes, thyroid problems, heart disease, rheumatoid arthritis and depression. So be sure to ask your doctor if they think any of these conditions may be contributing to your fatigue.

Managing Pain

To help your doctor prescribe the best medication, it's useful to give an accurate report of your pain. Keep a journal that includes information on:

- Where the pain occurs
- When the pain occurs
- How long it lasts

- How strong it is on a scale of 1 to 10, with 1 being the least amount of pain and 10 the most intense
- What makes the pain feel better and what makes it feel more intense

There are a number of options for pain relief, including prescription and over-the-counter medications. It's important to talk to a member of your health care team before taking any over-the-counter medication, to determine if they are safe and will not interfere with your treatments.

Physical therapy, acupuncture and massage may also be of help in managing your pain. Consult with a member of your health care team before beginning any of these activities.



Communicating With Your Health Care Team

As you manage your MPN, it's important to remember that you are a consumer of health care. The best way to make decisions about health care is to educate yourself about your diagnosis and get to know the members of your health care team, including doctors, nurses, nurse practitioners, physician assistants, dietitians, social workers and patient navigators.

Here are some tips for improving communication with your health care team:

Start a health care journal. Having a health care journal or notebook (either on paper or in a digital format) will allow you to keep all of your health information in one place. You may want to write down the names and contact information of the members of your health care team, as well as any questions for your doctor.

Prepare a list of questions. Before your next medical appointment, write down your questions and concerns. Because your doctor may have limited time, ask your most important questions first and be as specific as possible.

Bring someone with you to your appointments or have them be present during telehealth sessions. Even if you have a journal and a prepared list of questions or concerns, it's always helpful to have support during your appointments. The other person can serve as a second set of ears. They may also think of questions to ask your doctor or remember details about your symptoms or treatment that you may have forgotten.

Write down your doctor's answers. Taking notes will help you remember your doctor's responses, advice and instructions. You can also ask the person who accompanies you to take notes for you, either in your journal or on a tablet or smartphone.

Record your visit if your doctor allows it. Recording the conversation with your doctor gives you a chance to hear specific information again or share it with family members or friends.

Incorporate other health care professionals into your team.

Your MPN specialist (often a hematologist oncologist) is an essential member of your health care team, but there are other health care professionals who can help you manage your diagnosis and treatment:

- Your primary care physician should be kept updated about your MPN treatment and any test results.
- Your local pharmacist is a great source of knowledge about the medications you are taking. Have all of your prescriptions filled at the same pharmacy to avoid the possibility of harmful drug interactions.
- Make sure your oncologist knows of any other medical conditions you have or any pain you are experiencing so that they can consult with your primary care physician or specialists as needed.

Remember, there is no such thing as over-communication.



CancerCare's Free Support Services and Programs

It can be very difficult to receive a diagnosis of MPN, and adjusting to the necessary changes in your life can be challenging.

CancerCare® can help. We are a national nonprofit organization providing free, professional services to anyone affected by cancer. Our licensed oncology social workers can provide support and education, help in navigating the complicated health care system and offer information on support groups and other resources.

To learn more about how CancerCare helps, call us at 800-813-HOPE (4673) or visit www.cancercares.org.

You will likely also build your own personal support network composed of family and friends. In doing so, it's best to take some time to think about the people in your life and how they are best suited to help. Match the task to their strengths—ask a family member who loves to shop to pick up something for you at the store, or ask a friend who's a good listener to come over for a chat.



MORE ABOUT MYELOPROLIFERATIVE NEOPLASMS (MPNs)

Frequently Asked Questions

Q: Can an MPN progress to another condition?

A: In about 10-20 percent of cases, an MPN progresses to acute myeloid leukemia (AML), a cancer of the blood and bone marrow. Treatment options for AML depend on several factors, including its type, the results of lab tests and the person's overall health. Once AML is diagnosed, a treatment approach is decided, based on the person's individual circumstances. No matter what treatment approach is chosen, it is essential that people being treated for AML receive attentive supportive care. This care typically includes antibiotics to prevent or treat infections, which can occur due to the lower number of white blood cells seen in cases of AML. Supportive care also involves regular transfusions of red blood cells and platelets.

Q: I have been diagnosed with MF. Am I at higher risk of developing anemia?

A: People with MF can develop anemia due to a lowered production of red blood cells. Treatments for anemia include blood transfusions, an injection of the hormone epoetin alpha (Procrit) and immunomodulators such as thalidomide (Thalomid), lenalidomide (Revlimid) and pomalidomide (Pomalyst, Imnovid). Immunomodulators used to treat anemia in MF can cause fatigue, drowsiness, diarrhea, nausea and vomiting, loss of appetite and rash.

Q: What are the other types of MPNs?

A: Although polycythemia vera (PV), myelofibrosis (MF) and essential thrombocythemia (ET) are the primary types of MPN, there are other blood disorders that are classified as MPNs:

- **Chronic eosinophilic leukemia (CEL)** occurs when there is an abnormally high number of eosinophils (a type of white blood cell) in the blood. Treatment is typically determined by the cause of eosinophilia and is based on the symptoms being experienced.
- **Chronic myelogenous (CML)** involves an overproduction of granulocytes, a type of white blood cell. These cells accumulate, making it harder for bone marrow to make other blood cells the body needs.
- **Chronic neutrophilic leukemia (CNL)** occurs when too many neutrophils, a type of white blood cell, are made in the bone marrow. The typical treatment approach is symptom management and medications that slow the overproduction of white blood cells.
- **Myeloproliferative neoplasm, unclassifiable (MPN-U)** is a type of MPN that doesn't fit into any other category. It may involve an overproduction of various blood cell types, including white blood cells, red blood cells or platelets.

Q: Can you tell me more about telehealth?

A: Telehealth, also called telemedicine, refers to the remote delivery of health care services. Through video-conferencing technology, you can communicate with your doctors and other members of our health care team from the comfort of your home. The communication can also be via a traditional "voice only" phone call.

Through telehealth, members of your health care team can provide:

- A pre-visit review of your symptoms, personal health and family history
- A review of your treatment plan and expectations
- Guidance for in-person or hospital visits
- Information on relevant clinical trials
- Adherence guidance
- Symptom management (including pain)
- Education on lifestyle modification to improve your quality of life

Contact your health care team about whether telehealth appointments are available and if they are the right choice for you. Your insurance carrier can advise you about your coverage for telehealth appointments.

Q: What is a treatment summary and why is important?

A: Keeping your own records up-to-date in the form of a treatment summary can be helpful, as it allows you and your family members to have instant access to the specifics of your MPN diagnosis and treatment. A treatment summary should include:

- Your name and date of birth
- Date of diagnosis
- Prescribed therapy/therapies, including dates started and stopped and dosages when appropriate
- Dates and types of baseline and post-diagnosis testing and the results of these tests
- Other medications and supplements you are taking
- Names, affiliations and contact information of all members of your health care team

Take your personal record with you when you visit any doctor, not just the doctor treating your MPN.



Resources

CancerCare®

800-813-HOPE (800-813-4673)
www.cancercares.org

American Cancer Society

800-227-2345
www.cancer.org

Cancer.Net

Patient information from
the American Society of
Clinical Oncology
888-651-3038
www.cancer.net

National Cancer Institute

800-422-6237
www.cancer.gov

CLINICAL TRIALS WEBSITES

ClinicalTrials.gov

www.clinicaltrials.gov

EmergingMed

www.emergingmed.com

Cancer Support Community

888-793-9355
www.cancersupportcommunity.org

National Coalition for Cancer Survivorship

877-622-7937
www.canceradvocacy.org

MPN Research Foundation

773-977-7216
www.mpnresearchfoundation.org

MPN Education Foundation

www.mpninfo.org

Medicine Assistance Tool

www.medicineassistancetool.org

National Cancer Institute

www.cancer.gov

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