

FRANKLY
SPEAKING
ABOUT CANCER

Myelofibrosis



People with cancer who actively participate in their care along with their health care team will improve the quality of their lives and may enhance the possibility of recovery. People with cancer who actively participate in their care along with their health care team will improve the quality of their lives and may enhance the possibility of recovery.

Table of Contents

PROLOGUE	1
CHAPTER 1 / EMPOWER YOURSELF	2
CHAPTER 2 / UNDERSTANDING MYELOFIBROSIS	6
CHAPTER 3 / MYELOFIBROSIS SYMPTOMS & COMPLICATIONS	10
CHAPTER 4 / DIAGNOSIS & TREATMENT OF MYELOFIBROSIS.....	12
CHAPTER 5 / MANAGING SIDE EFFECTS	20
CHAPTER 6 / COPING WITH THE EMOTIONS OF MYELOFIBROSIS	24
CHAPTER 7 / MOVING FORWARD	27
CHAPTER 8 / GLOSSARY & RESOURCES	30

Prologue

This booklet is designed to help you take more control over your situation and your treatment decisions by giving you a better understanding of what **myelofibrosis** is and how it is diagnosed, what treatments are available, and what you may need to manage the emotional impact of **cancer**. It also provides information about credible organizations and resources that can help you find support, feel less alone, and enable you to develop a renewed sense of hope. Consider your healthcare team to be your partner throughout this journey.



I went to a doctor who treats MF patients. She gave me information and then I did my research. I am very proactive when it comes to that and I feel that you need to be. You have to have faith in your doctor but you definitely need to be proactive in continually researching what they give you.

— Olivia, myelofibrosis patient

1

Empower Yourself

THE IMPACT OF CANCER

Hearing the words “you have cancer” can be devastating to a person and their support system. Studies by the Cancer Support Community have identified three common emotional stressors that impact most people with cancer: an unwanted sense of loneliness, a loss of control, and a loss of hope. Emotional distress is common for people with cancer, and can include feelings of vulnerability, anger, sadness, depression, and anxiety.

In addition to emotional stress, there are also the practical elements of one’s life that are disrupted and may cause stress. These can include changes in family roles, a strain on financial resources, and a decrease in self-esteem.

Fortunately you can take steps to gain information, seek support, and learn how to overcome emotional distress to improve your quality of life.

THE EMPOWERMENT APPROACH

The Cancer Support Community utilizes the concept of empowerment to enable people affected by cancer to feel a greater sense of control over the disease.

This Empowerment Approach states that:

People with cancer who actively participate in their recovery along with their healthcare team will improve the quality of their lives and may enhance the possibility of their recovery.

People who adopt this philosophy:

- Make active choices in their treatment
- Make changes in their lives that they think are important
- Partner with their doctor through open communication
- Access resources and sources of support

Everyone's journey through cancer is unique. There is no instruction manual that tells you how to be empowered, and there's no right or wrong way to do it. Different people need different things. You decide what is best for you.



In a sense, slowing down is a positive thing. It's not all negative. I'm always aware of the gift of being able to appreciate what I do have and what I experience. You take less for granted.

— Jason, myelofibrosis patient

TEN ACTIONS YOU CAN TAKE TO IMPROVE YOUR QUALITY OF LIFE

1. **Take one day and make one decision at a time.**

Try to focus on resolving only today's problems. Avoid projecting worst-case scenarios for the future. Taking one small step at a time can help you feel less vulnerable and give you a greater level of control.

2. Ask for support. Be open with your family and friends about how you feel and how they can support you. Offer specific examples, such as: driving you to appointments, researching sources for financial support, or just listening when you want to talk. It is also good practice to take someone with you to medical appointments to take notes and help you remember instructions. Your nurse or oncology social worker can offer a wealth of information.

3. Communicate with your healthcare team.
Prepare a list of questions for each appointment. If you don't understand something, ask for clarification. Ask to see x-rays or scans to get a better picture of your status. If you do not develop a good relationship with your doctor, consider finding another. In general, it is useful to get a second opinion on your diagnosis and treatment to feel more confident about next steps.

4. **Retain as much control of your life as is**

reasonable. Having cancer can make it difficult for you to feel in charge of your life and your care. Work with your doctor, nurse, and caregiver to develop a plan that gives you as much control over your life as you desire and can comfortably handle.

5. Acknowledge and express your feelings. A diagnosis of cancer can trigger many strong emotions. Take time to listen to yourself. Find constructive ways to express your feelings through writing, talking, physical activity or creative pursuits. Consider professional support if you feel that depression or anxiety is hampering your ability to function well.

6. **Seek support from other cancer survivors.**

Often, people find a sense of comfort when they communicate with others affected by cancer. Ask your doctor, nurse, or social worker for local support groups. You can also contact the Cancer Support Community for ways to connect one-on-one, in groups, or online with others.

7. Learn relaxation techniques. "Relaxation" refers to a calm, controlled physical state that will enhance your well-being. Relaxation is something that you might have to learn, or build it into your

day, but it is important to do so. Consider music that makes you happy, reading a book, or taking a walk. Yoga, tai-chi, or meditation programs are also helpful. Take time to enjoy the moment.

- 8. Do what you enjoy.** Try to find humor in the unexpected moments of each day. Consider activities that you enjoy and can do comfortably. If you need to spend time alone, allow yourself that luxury.
- 9. Make healthy lifestyle choices.** It's never too late to make changes that will absolutely improve your well-being. Improving your diet, including exercise in your routine, and maintaining intimacy are all ways of feeling better both physically and emotionally.
- 10. Maintain a spirit of hope.** Hope is desirable and reasonable. There are millions of people who have fought cancer. Even if your cancer recovery is complicated, you can set small goals and enjoy daily pleasures. You may want to redefine how and when you experience hope by focusing on your spiritual beliefs, cultural customs, and family connections.

HELPING YOU PREPARE FOR CANCER TREATMENT DECISIONS



Open to Options™ is a free program designed to help:

- **Create** the list of questions you have for your doctor or healthcare team
- **Organize** your questions to bring with you to your appointment
- **Communicate** your questions and concerns clearly

IT WORKS! Patients who participated in the program:

- Were less anxious about their doctor appointment
- Felt that their appointment went more smoothly
- Felt better about the care decision made

It is easy and FREE to participate!

1-888-793-9355

www.cancersupportcommunity.org/

Open2Options

2

Understanding Myelofibrosis

UNDERSTANDING MYELOFIBROSIS

The Bone Marrow

In order to better understand myelofibrosis (MF), it helps to first have a basic understanding of the **bone marrow** and its many functions. The bone marrow is the spongy tissue inside the bones that houses red blood cells, white blood cells, and **platelets**. **Stem cells** are immature cells in the bone marrow that become these blood cells. Each of these cell types serves a critical function in the body. Red blood cells carry oxygen to tissues; white blood cells fight infections; and platelets help in clotting.

Myeloproliferative Neoplasms

Myelofibrosis is one of three most common diseases classified as a **myeloproliferative neoplasm** (MPN). MPNs are blood cancers that overproduce blood cells in the bone marrow, and are caused by genetic mutations in the blood stem cells. There are three main MPNs. The diagnosis depends on which of the blood cells are affected: Other myeloproliferative neoplasms include polycythemia vera (PV) and essential thrombocythemia (ET); in fact, ten to fifteen percent of myelofibrosis cases begin as either ET or PV.

Myelofibrosis

According to The Leukemia & Lymphoma Society's publication *Myelofibrosis Facts*, myelofibrosis occurs in about 1.5 out of every 100,000 people in the United States annually. The disease affects both men and women and is usually diagnosed in people over the age of 50; however, myelofibrosis can occur at any age. Myelofibrosis is a type of chronic leukemia (cancer of the blood) and can occur on its own (**primary myelofibrosis**) or as a result of another bone marrow disorder (**secondary myelofibrosis**). In primary myelofibrosis, a genetic mutation in a single hematopoietic (blood-forming) stem cell

DO I HAVE CANCER?

Because myelofibrosis was only recently classified as a cancer by the World Health Organization, there is often lack of consistent information offered to patients and caregivers when they are diagnosed. The Cancer Support Community conducted a survey in 2012 of MF patients to understand how patients see themselves and the challenges of a myelofibrosis diagnosis.

Only 14% of survey respondents were told they had cancer at the time of diagnosis and all of these respondents consider themselves to have cancer. Of those who were not initially told that they had cancer when they were diagnosed, 50% now consider themselves to have cancer; 14% still do not consider themselves to have cancer and 8% were not sure.

These results demonstrated the need for increased patient education and general disease awareness around myelofibrosis.

The results of this survey support the need for materials, like this book, to make sure patients have trusted and up-to-date information about myelofibrosis.

disrupts the body's production of normal blood cells, including the red blood cells, white blood cells, and platelets. As in most cancer processes, the abnormal cells begin to take over the normal cells, which leads to a disruption in the function that those cells serve. It also leads to extensive scarring in the bone marrow, known as fibrosis.

Myelofibrosis was classified as a neoplasm in 2010 by the World Health Organization (WHO). It was during this time period that people diagnosed with myelofibrosis were considered to have cancer. In people who have no symptoms, MF may be suspected when a routine medical checkup reveals an enlarged **spleen** and abnormal blood test results. The results of a blood test (a **complete blood count**, or CBC) that suggest a diagnosis of MF often include:

- A decrease in the number of **red blood cells** (**anemia**) below the normal range
- An increase or decrease in the number of **white blood cells**
- An increase in platelets above the normal range (occurs in about one-third of MF patients)
- A mild to moderate decrease in platelets below the normal range (occurs in about one-third of MF patients)

MYELOFIBROSIS FAST FACTS

- Myelofibrosis (MF) is a rare bone marrow cancer. It is one of a related group of blood cancers known as “myeloproliferative neoplasms (MPNs)” in which bone marrow cells that produce the blood cells develop and function abnormally. The resulting fibrous scar tissue formation leads to severe anemia, weakness, **fatigue** and an enlarged spleen and liver.
- MF is a type of chronic leukemia and can occur on its own (primary myelofibrosis) or as a result of another bone marrow disorder. Other MPNs that can progress to myelofibrosis include polycythemia vera and essential thrombocythemia.
- MF develops when a genetic mutation occurs in blood stem cells. The cause of the gene mutation is unknown. Between 50 and 60 percent of people with MF have a mutation of the Janus kinase 2 gene (*JAK2*). Researchers are investigating other possible gene mutations responsible for MF.
- MF usually develops slowly and some people may live symptom-free for years. Others, however, may get progressively worse, requiring treatment. In both cases, patients do need to be monitored regularly.
- The treatment goal for most patients with MF is to relieve symptoms and reduce the risk of complications. Treatments include blood **transfusions**, **chemotherapy**, radiation or removal of the spleen (**splenectomy**), drugs to treat anemia, and **allogeneic** stem cell transplantation. Ruxolitinib (Jakafi™) is the first drug approved by the US Food and Drug Administration (FDA) to treat MF because it has been shown to reduce several MF-related symptoms and control spleen enlargement. Other potential therapies for MF treatment are being studied in clinical trials.
- Allogeneic stem cell transplantation (ASCT) is the only potential cure for MF. This treatment has a considerable risk of life-threatening side effects. ASCT is not a good option for most people with MF because of age, the course of their disease or other health problems.

Excerpt from The Leukemia & Lymphoma Society's *Myelofibrosis Facts*

3

Myelofibrosis Symptoms & Complications

Myelofibrosis typically develops very slowly and, therefore, symptoms do not commonly develop early in the disease process. However, as the abnormal cells increase, they cause more and more disruption in the functioning of the normal blood cells.

SYMPTOMS OF MYELOFIBROSIS

- Fatigue, weakness, or shortness of breath. These symptoms are associated with a decrease in the red blood cells (anemia)
- Pain or fullness below the ribs on the left side, as a result of an enlarged spleen (**splenomegaly**)
- Enlarged liver (**hepatomegaly**)
- Pale skin
- Easy bruising or bleeding, due to a decrease in the platelet count (**thrombocytopenia**)
- Flat, red, pinpoint spots under the skin caused by bleeding (**petechiae**)
- Excessive **night sweats**
- Fever
- Frequent infections, due to a low white blood cell count (**neutropenia**)
- Bone or joint pain
- Weight loss
- Itching (**pruritus**)

COMPLICATIONS FROM FURTHER PROGRESSION OF MYELOFIBROSIS

Portal hypertension

This is an increase in the blood pressure of the portal vein, which is the vein that carries blood from the spleen to the liver. When the volume of blood in the spleen increases, so does the pressure in the portal vein. This may force excess blood into smaller veins in the stomach and esophagus, possibly causing the veins to rupture and bleed.

Extramedullary hematopoiesis

This is when a formation of blood cells outside the bone marrow creates clumps, or tumors, of developing blood cells in other areas of your body. These tumors may lead to bleeding in your gastrointestinal system, coughing or spitting up of blood, compression of your spinal cord, or seizures.

Hardening and inflammation of bone tissue

Myelofibrosis can cause hardening of the bone marrow and inflammation of the connective tissue that surrounds the bones. This results in severe bone and joint pain and tenderness.

Gout

This is a condition caused by an overproduction of uric acid. Because of this overproduction, needle-like deposits develop in the joints, causing joint pain and inflammation.

Acute leukemia

Some people with myelofibrosis eventually develop acute myeloid leukemia, a type of blood and bone marrow cancer that progresses rapidly.

Just trying to get the diagnosis of myelofibrosis for my wife was a challenge. She underwent so many tests, and didn't understand at first if it was even cancer. However, once we finally got the right diagnosis, we could form a plan and start treatment.

— Myelofibrosis Caregiver

4

Diagnosis & Treatment of Myelofibrosis

Most commonly, people diagnosed with myelofibrosis do not have obvious symptoms. MF may be suspected when an enlarged spleen or an abnormal blood count is noted upon a routine checkup. At that point, your general practitioner will likely refer you to a hematologist/oncologist (a doctor who specializes in blood cancers). Your **hematologist/oncologist** will coordinate your treatment plan, so it is important that you find someone in this field that you feel comfortable working with. For more information about choosing a doctor or a treatment center, see the free publication *Choosing a Blood Cancer Specialist* or *Treatment Center* published by The Leukemia and Lymphoma Society.



The spectrum of treatment for myelofibrosis can be overwhelming at first. Although there are a variety of treatment approaches, you and your doctor will need to have ongoing discussions about how your disease can best be managed over time.

— Ross Levine, MD, Memorial Sloan Kettering Cancer Center

WHEN TO GET A SECOND OPINION

It is always a good idea to consider a second opinion. Getting a second opinion can provide you with additional information, treatment options, or access to a different medical facility. It may also give you confidence that you are already on the right course. Some insurance companies require a second opinion and others will cover the cost if you request one.

Other reasons to consider a second opinion:

- If you feel uncertain about your diagnosis or treatment options.
- If you were diagnosed by someone who has limited experience in treating myelofibrosis and would like to speak with someone who has more experience.
- If you are interested in a clinical trial.

People should know about other doctors offering other treatments. Get more opinions; make your own treatment plan. I haven't found a doctor to lead my treatment. I lead my treatment.

— *Myelofibrosis Patient*

FINDING AN EXPERIENCED DOCTOR

It is very important to find a doctor who is experienced with treating myelofibrosis, who answers your questions, and makes you feel comfortable.

Never be afraid to ask your doctor important questions such as:

- How much experience do you have in treating myelofibrosis?
- Are you board-certified in this specialty?
- Do you stay up-to-date with the latest treatments for myelofibrosis? If so, how?
- Do the doctors at your clinic/hospital participate in clinical trials?
- Do you accept my insurance (cost of care)?
- Will you refer me to an oncology nurse and/or social worker to help me with education, resources, and support?
- What other support services are available to me and my family?
- How often and when can I contact my treatment team with questions? (Can I use email or phone, or do you prefer only office visits?)

WHAT DIAGNOSTIC TESTS SHOULD YOU EXPECT?

A combination of tests that examine the blood and bone marrow will be used to determine the course of treatment.

In addition to a physical exam and history, the following diagnostic tests may be used:

A **complete blood count**, also known as a CBC, might show one or more of the following results that might suggest a myelofibrosis diagnosis:

- Decreased red blood cell count (anemia)
- Increased white blood cell count
- Either an increased OR decreased platelet count

In addition to the blood cell counts being out of normal range, there may be abnormalities noted in the shape, size, or maturity of the cells that could indicate a problem. There may also be certain levels in the blood that are either elevated or low, which may also prompt further testing.

- **Ultrasound** takes a picture called a sonogram created by bouncing sound waves off organs in the abdominal cavity, to see if anything abnormal exists. This would be used to see if there are any abnormalities in the spleen or liver.

- **Magnetic Resonance Imaging (MRI)** can provide a detailed picture of the organs and bones in the body, created by using a magnet, radio waves and a computer. This would be used to identify changes in the bones that indicate myelofibrosis.
- **Bone Marrow Examination** can confirm a myelofibrosis diagnosis. By removing a small amount of your bone and bone marrow and studying them under a microscope, genetic components can be seen that can help in both diagnosis and treatment decisions. These are usually done at the same time as an outpatient procedure at the hospital.
 - The aspiration removes the liquid portion of bone marrow.
 - The biopsy and aspiration are usually done at the same time as an outpatient procedure.
- **Genetic tests** may also be done by analyzing a sample of your blood or bone marrow tissue. Certain genetic mutations, such as the *JAK2* mutation, are seen in the majority of myelofibrosis patients.

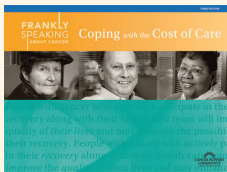
TREATMENT OF MYELOFIBROSIS

Who is involved in the treatment of myelofibrosis?

There may be several doctors involved in your care. In addition to your hematologist/oncologist you might also have a surgeon, radiation oncologist, and/or a stem cell transplant specialist.

Other key players on your medical team include support persons, such as your oncology nurse, nurse navigator, oncology social worker and pharmacist. Each of these professionals can help guide you to information and resources that may help you both during and after your treatment. If you do not meet these professionals upfront, ask your doctor how you might arrange an appointment with them.

In addition, you may want to contact your insurance carrier to ask for a case manager to be assigned to you to assist in managing your treatment coverage options.



For more information on the cost of care, please refer to CSC's **Frankly Speaking About Cancer: Coping with the Cost of Care** at 1-888-793-9355 or www.cancersupportcommunity.org.

APPROACHES TO TREATMENT

Watch and Wait

Patients who are symptom free and do not have signs of anemia, an enlarged spleen or other complications are generally not treated at the time of diagnosis. Some people remain stable and symptom free for many years. However, it is very important that such patients be closely monitored through regular medical checkups and examinations to detect any signs or symptoms of disease progression.

Drug Therapies

There are various drugs used in the treatment of myelofibrosis, and many of them have very different approaches to slowing the disease process and/or controlling the effects of the disease. With any drugs, side effects will likely exist; your doctor or nurse should review these with you before starting.

- **Chemotherapy** — These are drugs that kill cancer cells that divide rapidly. These agents also attack other fast growing cells in the body, such as nails and hair and may cause other more serious side effects. These drugs may be given by mouth (orally) or into the vein (intravenously).

- **Janus-associated kinase (JAK) inhibitor** – This class of drug inhibits enzymes called “JAK1” and “JAK2”, which are involved in regulating blood. It is important to note that JAK inhibitors work in both patients that have a JAK mutation and those that do not have a JAK mutation. The JAK inhibitor is the first FDA approved therapy for patients with MF. Their focus is to treat and lessen some of the symptoms of MF, including an enlarged spleen, night sweats, itching, and bone or muscle pain. An increased risk of bleeding and infection may occur as may bruising, dizziness and headaches.
- **Immunomodulators (IMiDs)** – The drugs in this class work against cancer cells by affecting the functions of the immune system. Certain immunomodulators are used to treat MF patients for anemia. Another immunomodulator has been used to treat an enlarged spleen, bone pain and high platelet count in selected MF patients. Other immunomodulators are under study in clinical trials.
- **Androgen therapy** – These drugs are synthetic forms of male hormones, and can promote red blood cell production. They are used to relieve the symptoms of severe anemia in MF patients. These drugs can be toxic to the liver, and, therefore, require close monitoring of liver functions through blood tests and ultrasound. They may also cause facial hair growth and other masculinizing effects in women.
- **Recombinant erythropoietin** – This treatment helps regulate red blood cell production. It is given by intramuscular (into the muscle) or subcutaneous (under the skin) injection. However, the response of anemia-related symptoms in MF patients has been limited.
- **Glucocorticoids** – Also, known as “steroids,” these drugs are used to treat various conditions. They are used in MF patients to treat significant anemia. A number of side effects may occur when using glucocorticoids but are usually manageable with short term use.
- **Bisphosphonates** – These drugs are used to prevent bone loss in cancer patients, and may relieve bone pain, as well as increase blood counts in MF patients. These drugs are given intravenously. Short term use may cause upset stomach.
- **Anagrelide hydrochloride** – This drug, given orally, may be used in MF patients who have an extremely high platelet count, especially following a splenectomy. Headache, diarrhea, weakness, nausea and dizziness may occur.

Radiation Therapy

Radiation uses high-powered beams, such as X-rays, to kill cells. **Radiation therapy** can help reduce the size of the spleen, when surgical removal isn't an option. It is also used to treat bone pain and tumors outside the bone marrow.

Surgical Removal of the Spleen (Splenectomy)

If your spleen is enlarged and causing harmful complications and pain, and other interventions have not been successful, you may benefit from having your spleen surgically removed. Your doctor will consider the risks and benefits of your particular situation and determine if this is a feasible option. If surgery is done, you should be closely monitored for complications.

Stem Cell Transplantation

This is the only type of treatment that has the potential to cure myelofibrosis. However, it is a very high risk treatment, and can only be considered for patients deemed appropriate, and is dependent on the availability of a suitable donor.

- **Allogeneic stem cell transplant** – In this procedure, the patient receives high doses of chemotherapy or radiation to destroy the diseased bone marrow. The donor stem cells (from someone other than the patient) are then infused into the patient.
- **Non-myeloblastic stem cell transplant** – This transplant uses lower doses of chemotherapy or radiation prior to the infusion of donor cells. This type of transplant can be used in older and sicker patients, who may not be eligible for an allogeneic transplant.

Clinical Trials

Because doctors are still trying to find more effective treatments for myelofibrosis, clinical trials are an important option to consider. Clinical trials are research studies to test promising new drugs or treatment approaches. Participating in a clinical trial may give you the opportunity to be among the first to benefit from the most advanced treatment available. If you are interested in a clinical trial, ask your healthcare team about studies that you might qualify for.

The latest research in myelofibrosis involves looking at the effectiveness of several different JAK-2 inhibitors in patients with MF. This treatment stems from the knowledge that the mutation of the JAK2 gene is one mutation that may lead to the development of myelofibrosis. JAK2 inhibitor drugs are used for all mutations, including those who test JAK2 negative. Some of the benefits of such therapies may include: reducing spleen size, improving anemia, platelet count, and other symptoms, including fatigue, night sweats, weakness, and shortness of breath.

TYPES OF CLINICAL TRIALS	
TYPE OF TRIAL	STUDY GOALS
Prevention trials	What can prevent cancer from developing?
Screening and early detection trials	What new screening and detection methods can be used to identify cancer in people, before it spreads and before symptoms?
Diagnostic trials	How can new tests identify a cancer earlier and more accurately?
Treatment (or therapeutic) trials	What new treatments can be used to stop or cure cancer?
Quality of life and supportive care trials	What can be done to improve the overall well-being of people who have cancer? What can be done to make people who have cancer more comfortable?

5

Managing Side Effects

Just as each person's treatment plan is individualized, the side effects that each person experiences might be very different among patients. For this reason, we provide only general information here and we encourage you to work closely with your healthcare team to manage any side effects that **you** experience.

The side effects of cancer treatment are due to the specific type of treatment(s) that you receive. They can be mild to severe. It helps to learn about potential side effects before you begin. Ask any and all questions that you might have so you can work with your healthcare team to manage your quality of life during and after treatment. Fortunately, there are very effective and readily available medications to address traditional side effects (such as nausea, diarrhea, constipation, fatigue, low blood counts and mouth sores), as well as side effects from targeted treatments.

MANAGING ANEMIA

Everyone responds differently to treatment. Many variables affect a person's response including:

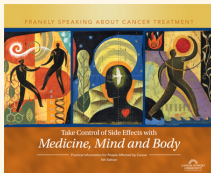
- type of treatment and dosage
- the age of the patient
- coexisting medical conditions

While no one can predict with certainty how you will respond to a particular treatment, your healthcare team is there to help manage the side effects that you experience. It is critical, therefore, that you communicate how you're feeling and any concerns you may have with your doctor and/or nurse.

Anemia is a very common effect of myelofibrosis and there are many treatment approaches to improve red blood cell counts in patients with myelofibrosis. As mentioned in the treatment section of this booklet, there are many drug therapies targeted at improving anemia in patients. Blood transfusions are also an option for patients when their red blood cell count is very low.

There are additional non-drug therapies that can assist in managing your anemia. Because anemia may also cause fatigue and shortness of breath, there are a number of things you can do to conserve energy including pacing your activities, taking frequent breaks, getting plenty of rest and maintaining good nutrition.

LEARN MORE ABOUT SIDE EFFECT MANAGEMENT



Useful tips to better manage or possibly prevent common side effects such as fatigue, nausea, gastrointestinal problems, skin rashes, low blood counts and hair loss can be found in the **Cancer Support Community's** booklet: ***Frankly Speaking About Cancer Treatment: Take Control of Side Effects with Medicine, Mind, and Body*** (order a free booklet or view <http://cancersupportcommunity.org/fsacmaterials>).

SIDE EFFECTS OF SURGERY (SPLENECTOMY)

Your body will need time to recover from surgery. Initially, you will have pain or discomfort and medication to control the pain will be ordered for you after surgery. You will be asked to rate your pain on a scale of 0 (no pain) to 10 (very painful). Be sure to communicate openly to your nurse or doctor about your pain and let your healthcare team know if your pain is not well controlled.

Because pain killers can slow the movement of the bowel, your healthcare team will monitor this closely. You may need to take medications temporarily to help return your bowel function to normal.

- The incision from your surgery will take time to heal. Your nurse or a homecare nurse will teach you how to take care of the incision when you return home. If you notice any redness or pus at the surgical site or if you develop a fever, contact your doctor right away. Following splenectomy, people have increased risk of severe infections, especially during the first two years after surgery.
- You may feel tired or weak after surgery. Take short naps (20-30 minutes), prioritize the things that are most important for you to accomplish and delegate the rest of your tasks to family or friends.

SIDE EFFECTS OF DRUG THERAPIES

Every drug causes a unique set of side effects. Your oncology nurse will discuss the side effects you can expect based on your treatment plan. If you are taking a drug as part of a clinical trial, the research nurse will discuss any side effects you should anticipate for the specific drug you are taking. Ask for written information about the drugs you are taking and the best way to manage those side effects.

SIDE EFFECTS OF STEM CELL TRANSPLANT

Once a donor's stem cells are infused, there is a risk that the new stem cells will react against your body's healthy tissues, causing potentially fatal damage (graft-versus-host disease). Other risks include organ or blood vessel damage, cataracts and the development of a different cancer later on.

SIDE EFFECTS OF RADIATION THERAPY

If you receive external radiation therapy, you may experience nausea, vomiting or diarrhea. Radiation also causes fatigue and can reduce your blood count, leading to:

- Low white blood cells, which increases your risk for infection
- Low platelets, which increases your risk for bleeding
- Low red blood cells, which may make you feel tired or short of breath

Radiation therapy can also cause skin changes that include redness, dry or peeling skin, itching, blisters, and swelling.

- Your radiation oncologist and nurse will talk to you about skin care. Be gentle with your skin. Don't use any lotions or creams unless the radiation treatment team recommends them.
- Don't put anything hot, like a heating pad, or cold, like an ice pack, on the area being treated.
- Wear soft clothes and use soft sheets made of fabrics like cotton.

Call your nurse or doctor if your skin blisters or swells, you have diarrhea for more than 24 hours, or you are too nauseated to drink liquids after receiving external radiation therapy. Let your healthcare team know if you have a fever or any signs of infection, bleeding or are short of breath.

6 *Coping with the Emotions of Myelofibrosis*

Your myelofibrosis diagnosis affects the way that you feel, think and act. Just as you need to take care of your body, you need to take care of your emotions.

Common emotions you may experience:

- Anxiety
- Depression
- Anger
- Irritability
- Grief and loss for your old life (“loss of normal”)
- Helplessness
- Frustration
- Fear

One thing I have found is that it helps me to talk about my diagnosis, especially when family and friends take the time to ask and try to understand this disease. Knowing that people care and want to know what I am going through gives me strength and hope.

— Dorothy, myelofibrosis patient

The ways people deal with emotions are often referred to as coping mechanisms. Every person uses different ways to manage stress, depression, worry about what the future holds or other emotions. Learn how to effectively manage your emotions. This can help improve many aspects of your life – eating, sleeping, working, relationships with others and enjoyment of leisure activities. There are many resources available to assist you in managing your emotions should you decide to seek help. We encourage you to contact the Cancer Support Community Helpline at 888-793-9355 for further information and help in finding resources to help you cope with your diagnosis or the diagnosis of a loved one.

**CANCER SUPPORT COMMUNITY
HELPLINE: 1-888-793-9355**

CSC's Helpline is staffed Monday through Friday from 9 am to 8 pm EST by licensed mental health professionals. All services are at no cost to callers. Counselors can provide information, resource referral, psychosocial support, access to Open to Options treatment decision support, referrals to CSC programs, affiliates and online groups.

**TIPS TO HELP YOU MANAGE
FEAR AND ANXIETY**

- Educate yourself - learn what physical and mental signs or symptoms you should be aware of
- Keep up with any medical tests or appointments your doctor recommends
- Express your feelings with others who can understand your feelings either in a support group, with family and friends or a counselor
- Meditate or do deep breathing and relaxation exercises
- Keep a journal and write down your thoughts
- Take control of what you can, make a plan, and know what you cannot control
- When you have health-related questions or concerns write them down and bring to your appointments or call your healthcare team if you have immediate concerns

GENERAL STRATEGIES FOR WELLNESS

Take good care of yourself while you are undergoing cancer treatment:

- Good nutrition maintains energy, strengthens the immune system, can decrease side effects, and provides comfort.
- Adequate fluid intake avoids dehydration, prevents constipation, and decreases fatigue.
- Try to get some physical activity every day. Exercise combats fatigue, helps maintain physical function, promotes restful sleep, and can elevate your mood.
- Good personal hygiene decreases the risk of infection, keeps your skin healthy, and enhances your feelings of wellness.
- Regular sleep is important. If you are having trouble sleeping you should bring it to the attention of your healthcare team.
- Mind-body activities such as yoga, meditation and guided imagery help to reduce stress and increase energy.
- Participation in a support group can help you understand and manage your feelings and provide an opportunity for you to connect with others facing cancer.
- If you have questions about activities you may or may not participate in, be sure to ask your healthcare team.
- Being empowered in your cancer journey means asking questions, being informed, and making choices and decisions about your treatment and care that are right for **you**.

7

Moving Forward

LIFE WITH A CHRONIC ILLNESS

Myelofibrosis is not always curable, and for this reason, it is necessary to consider that the changes that lie ahead may not be temporary, but possibly lifelong. When you have a chronic illness, it becomes a part of who you are and how you live your life. It may require changes in your routine or in how you care for yourself, as well as what you need from others in your life. It does not, however, define you. You are not your disease.

You will need to be monitored on a regular basis. Your healthcare team will likely become individuals who you value as “key players” in the management of your illness. How frequently you see your healthcare team will depend on many factors, and will need to be an ongoing discussion between you and your doctor.



A diagnosis of myelofibrosis can feel overwhelming at first, but there is hope, you can regain some control in your life and you can find that ‘new normal’ as you realize you are not alone.

— *Melissa Wright, LMSW, OSW-C, Gilda's Club Quad Cities*

POSITIVE LIFE CHANGES

Instead of returning to normal, you will likely have to search for a “new normal.” Some cancer survivors talk about the “gifts of illness” – positive changes that occur as an outcome of a trauma or a life crisis.

Because cancer exposes you to the fragility of life, it gives you an opportunity to do things you have left undone. You may want to tell family members or friends that you love them, say thank you, tell someone that you forgive them or ask for their forgiveness.

CANCER MAY:

- Let you feel a greater sense of self-esteem or mastery
- Help you recognize the importance of surviving, find new meanings for life, and acquire stronger coping skills
- Enable you to develop greater insights into what is important to you
- Give you a desire to “give back”
- Show you a different orientation to time—living in the present to do things now rather than later
- Let you feel a greater appreciation for life

MAKING PLANS

It is a good idea for all adults, no matter their health situation, to prepare for the future by having their affairs in order. The following legal documents are tools to help you do that:

- A **will** is a legal document that designates who will receive your money and belongings and who will be responsible for your children (in the absence of another parent). If there is no will in place, an agent of the state may become responsible for these decisions.
- A **living will** is a legal document to let your doctor know what kind of medical care you want at the end of your life if you can't speak for yourself.
- A **durable power of attorney** is a person who will make healthcare decisions for you if you are unable to make them yourself.
- A **do not resuscitate (DNR)** order is an order for your doctor if you decide that you would not like heroic measures to be taken in the event of cardiac or respiratory arrest.

Choose someone you feel very comfortable with to assist you in making plans like these. If you make these plans, you will help your family and loved ones by not leaving them with terribly difficult decisions. You will be reducing confusion and helping everyone around you feel more comfortable that the decisions made are yours.



Through the support groups, these are people that I would have never met if I didn't have this disease and joining these groups. I have met wonderful people who will be lifelong friends. We have this common bond. People think that support group meetings are so depressing, but it is not like that. Knowing that people care and want to know what I am going through gives me strength. I know people care and the support is what gives me strength.

— Oliva, myelofibrosis survivor

FINDING HOPE

People affected by cancer hope for many things: peace in relationships; good communication with doctors; a way to come to terms with cancer; and certainly for the recovery of physical and emotional health.

By being empowered during cancer treatment and with your support systems, you can balance optimism and hope with a realistic understanding of your experiences. You can more effectively deal with information, regain a sense of control, and find hope that you are not alone.

Hope is found throughout the cancer experience in a variety of ways – and science is offering new hope for the best possible treatments available today and in the future.

8

Glossary & Resources

Allogeneic – taken from different individuals of the same species

Anemia – a condition in which the number of red blood cells in the body is below normal. Anemia can cause fatigue, weakness, and shortness of breath.

Bone Marrow – a tissue located inside the bones that makes blood cells

Cancer – a term for diseases in which abnormal cells divide without control and can invade nearby tissues. Cancer cells can also spread to other parts of the body through the blood and lymph system.

Complete Blood Count (CBC) – a blood test that measures the concentration of white blood cells, red blood cells, and platelets in the blood

Chemotherapy – treatment drugs that attack rapidly dividing cancer cells

Fatigue – a condition marked by extreme tiredness and inability to function due to lack of energy

Fibrosis – extra tissue that forms in the bone marrow that can affect its ability to function

Genes – the basic building blocks of heredity that are present in all cells

Hemoglobin – the part of the red blood cell that carries oxygen

Hematologist/Oncologist – a doctor who specializes in cancers of the blood

Hematopoietic stem cell – stem cells that give rise to all the various types of cells in the blood

Hepatomegaly – enlargement of the liver

MRI scan – a scan that uses magnets and radio frequency waves to produce images of the inside of the body

Myelofibrosis – a blood cancer in which the bone marrow is replaced by fibrous tissue and is no longer able to produce adequate numbers of normal blood cells

Myeloproliferative Neoplasms – a group of blood cancers - including **myelofibrosis**, **polycythemia vera** and **essential thrombocythemia** – in which large numbers of abnormal red blood cells, white blood cells or platelets grow and spread in the bone marrow and the blood

Neoplasm – an abnormal mass of tissue that results when cells divide more than they should or do not die when they should. Neoplasms may be benign (not cancer), or malignant (cancer). Also called tumor.

Neutropenia – a significant decrease in the number of white blood cells in the blood

Night sweats – episodes of excessive nighttime sweating. These can occur even if the room is not warm.

Petechiae – flat, red, pinpoint spots under the skin caused by bleeding

Platelets – small cell fragments that help blood to clot

Primary Myelofibrosis – myelofibrosis that occurs on its own, due to a genetic mutation

Pruritus – severe itching

Radiation therapy – a type of treatment that uses high energy to kill cancer cells

Red Blood Cells (RBCs) – cells that carry oxygen through the body

Risk factors – anything that increases the chance of getting a certain disease

Secondary Myelofibrosis – myelofibrosis that occurs as a result of another bone marrow disorder (usually PV or ET)

Spleen – an organ that is part of the lymphatic system. The spleen makes lymphocytes, filters the blood, stores blood cells, and destroys old blood cells. It is located on the left side of the abdomen near the stomach.

Splenectomy – surgical removal of the spleen

Splenomegaly – enlargement of the spleen

Stem Cell – a cell that can become another more mature type of blood cell

Thrombocytopenia – a significant decrease in the number of platelets in the blood

Transfusion – a procedure in which a person is given an infusion of blood. The blood may be donated by another person, or it may have been taken from the patient earlier and stored until needed. Also called blood transfusion.

Ultrasound – uses high-frequency sound waves to look at organs and structures inside the body

White Blood Cells (WBCs) – blood cells that help fight infection

RESOURCES

Cancer Support Community

www.cancersupportcommunity.org

HelpLine 888-793-9355

American Cancer Society

www.cancer.org

800-227-2345

Cancer Care

www.cancercare.org

800-813-4673

Chronic Disease Fund

www.cdfund.org

The Leukemia & Lymphoma Society

www.lls.org

800-955-4572

MPN Education Foundation

www.mpninfo.org

MPN Research Foundation

www.mpnresearchfoundation.org

National Cancer Institute

www.cancer.gov

800-422-7237

National Organization of Rare Disorders

www.rarediseases.org

CONTRIBUTORS

Ann Brazeau

MPN Research Foundation

Antje Hjerpe

MPN Education Foundation

Elizabeth Kitlas, LMSW

The Leukemia & Lymphoma Society

Ross Levine, MD

Memorial Sloan Kettering
Cancer Center

Yvette Payne APRN, MSN, MBA

Independent Oncology Consultant

**Lisa Marquette Porat, MSW,
LCSW, OSW-C**

Laura Weiger, MSW, LCSW

Melissa Wright, LMSW, OSW-C
Gilda's Club Quad Cities

DESIGN

Suzanne Kleinwaks Design, LLC

REFERENCES

All sources used for this publication are from credible, peer reviewed medical journals. If you'd like more information about specific sources, please contact the Education Department of the Cancer Support Community.

THE CANCER SUPPORT COMMUNITY (CSC)

www.cancersupportcommunity.org

CSC offers a wide range of support services to all people affected by cancer based on evidence that the best cancer care includes emotional and social support. As the largest professionally-led network of cancer support worldwide, we deliver personalized services including: support groups; one-on-one counseling; telephone support counseling; exercise; nutrition; and more programs at no cost.

THE LEUKEMIA & LYMPHOMA SOCIETY (LLS)

www.lls.org

LLS is the world's largest voluntary health organization dedicated to funding blood cancer research, education and patient services. LLS has chapters throughout the country and in Canada. Information Specialists, who are master's level oncology professionals, are available to help cancer survivors navigate the best route from diagnosis through treatment, clinical trials and survivorship.

MPN EDUCATION FOUNDATION

www.mpninfo.org

The MPN Education Foundation provides information, education and support and looks to advance research to improve the quality of life and care of MPN patients. The Foundation provides patient and doctor conferences and facilitates patient participation and accrual in clinical studies and surveys.

THE MPN RESEARCH FOUNDATION

www.mpd-rc.org

The MPN Foundation is a nonprofit organization whose primary mission is to promote, fund and support the most innovative and effective research into the causes, treatments and potentially the cure for essential thrombocythemia, polycythemia vera and MF.



The Cancer Support Community, and its partners, The Leukemia & Lymphoma Society, MPN Education Foundation and MPN Research Foundation, provides this information as a service. This publication is not intended to take the place of medical care or the advice of your doctor. We strongly suggest consulting your doctor or other healthcare professional to answer questions and learn more.

© 2012 Cancer Support Community. All rights reserved.

THIS PROGRAM WAS MADE POSSIBLE WITH A CHARITABLE CONTRIBUTION FROM:



People with cancer who actively participate in their care along with their health care team will improve the quality of their lives and may enhance the possibility of recovery. People with cancer who actively participate in their care along with their health care team will improve the quality of their lives and may enhance the possibility of recovery.

