LEUKEMIA & LYMPHOMA SOCIETY OF CANADA®

Myelofibrosis
MF

WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with myelofibrosis (MF). What does it mean and how will it affect you?

This fact sheet will help you:

- Learn about MF and how it is diagnosed
- Get an overview of treatment options
- Understand what happens next

What is myelofibrosis (MF)?

MF is a rare type of bone marrow cancer. It is a form of myeloproliferative neoplasm (MPN), where the bone marrow makes too many of one kind of blood cell. MPNs begin with one or more changes to the DNA of a single stem cell in the bone marrow. These changes cause the stem cell to create more and more abnormal stem cells.

In MF the abnormal cells cause normal bone marrow to be replaced by scar tissue. Over time, the fibrous scar tissue makes it harder for the bone marrow to produce normal blood cells.

About MF

- MF often develops slowly, and you may not have symptoms for years.
- Primary MF means it is your first MPN; secondary MF is when it developed from another MPN.
- For 10% to 20% of people with MF, the disease may progress to a more aggressive blood cancer, like acute myeloid leukemia.

Signs and symptoms

MF is often detected during a routine blood test. As your bone marrow produces more abnormal blood cells, you may experience:

- Fatigue, weakness, shortness of breath, and pale skin
  - When your red blood cell count is low (anemia)
- Feeling of fullness, abdominal pain, and decreased appetite
  - When you have an enlarged spleen or liver
- Excessive bleeding or bruising
  - When you have a low platelet count
- Night sweats and itchy skin
  - Possibly a response from your immune system
- Infections and fever
  - When your white blood cell count is low
- Bone or joint pain
  - When your white blood cells build up, causing your bone marrow to expand
- Weight loss
  - When you are eating less or using more energy

After your diagnosis

With your diagnosis, your doctor can determine the right treatment for you. Your test results help your doctor predict how MF will likely progress and how you may respond to treatment.

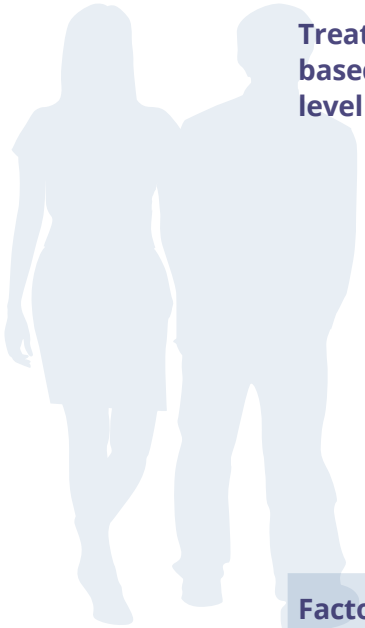
Name of test	Description
Medical history and physical exam	The doctor reviews past illnesses, injuries, and symptoms. They examine your lungs, heart, and other organs.
Complete blood count (CBC)	This test measures the number of red blood cells, white blood cells, and platelets in your blood. In people with MF, red blood cell levels are often low.
Peripheral blood smear	This test looks at blood cells under a microscope to see the number, size, shape, type, and pattern of cells. It also looks for blast cells, which healthy people do not usually have. People with MF often have abnormal teardrop-shaped red blood cells and blast cells.
Comprehensive metabolic panel	This group of blood tests, or blood chemistry profile, measures the levels of certain substances released into your blood.
Bone marrow aspiration and biopsy	These two tests examine bone marrow cells and look for scar tissue (fibrosis) and abnormal cells. They are usually done at the same time.
Molecular testing	This genetic test looks for a specific mutation of three genes (JAK2, CALR, and MPL) when MF is suspected.
Imaging tests	An ultrasound test is used to assess your spleen and liver.

MF treatment

Some people with MF will have no symptoms for years. There is not one specific treatment for everyone with MF. Different symptoms call for different treatments. Your treatment will focus on controlling symptoms and complications. It also aims to improve your quality of life. It's important for you to be monitored closely.

Types of treatment

- **Drug therapy** is the main form of treatment. It may include:
  - **Ruxolitinib** reduces the spleen size and treats MF symptoms.
  - **Hydroxyurea** decreases the number of blood cells made in your bone marrow.
- **Allogeneic stem cell transplantation** transfers a healthy person's (donor) stem cells to your body. This can be a high-risk procedure. For people with MF, it may be the only option to try to cure the disease, but for most, it's too risky.



Treatment based on risk level

Scoring systems are available to estimate prognosis in MF based on your blood counts, age, and symptoms. Your risk score can help your doctor recommend the best treatment for your disease.

- **Low risk with no symptoms** – You will be closely monitored, with regular checkups and examinations. No medication is needed.
- **Low risk with symptoms** – You may be prescribed ruxolitinib or interferon alpha.
- **Intermediate 1 risk** – Treatment may include ruxolitinib or allogeneic stem cell transplantation, if you are eligible.
- **Intermediate 2 and high risk** – Treatment may include ruxolitinib or allogeneic stem cell transplantation, if you are eligible.

Factors that affect treatment

Discuss your treatment options with your doctor to make sure you understand the benefits and risks of each approach. Your treatment plan is based on your age at the time of treatment, symptoms, and risk level.

Treatment side effects

When you begin your treatment for MF, you may experience mild to severe side effects, depending on your age, your overall health, and your treatment plan. New drugs and therapies can help control side effects. Speak to your doctor if you are having side effects.

Common side effects	You may experience side effects such as: • Fatigue, bruising, and bleeding from ruxolitinib treatments • Mouth ulcers (open sores), changes in your sense of taste, skin ulcers, or rash from hydroxyurea treatments • A slight increase in the risk of infections from ruxolitinib treatments
Living with MF	Medical follow-up is important with MF. Your medical team should provide you with a care plan listing the frequency of follow-up visits and the tests you will have at those visits.

Living with MF can be overwhelming. Seek medical help if you are feeling "down" or "blue" or don't want to do anything – and your mood does not improve over time. These could be signs of depression, an illness that should be treated even when you're undergoing treatment for MF. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

This fact sheet was reviewed by:

Dr Lynda Foltz, MD, FRCPC
St. Paul's Hospital
Clinical Professor
University of British Columbia