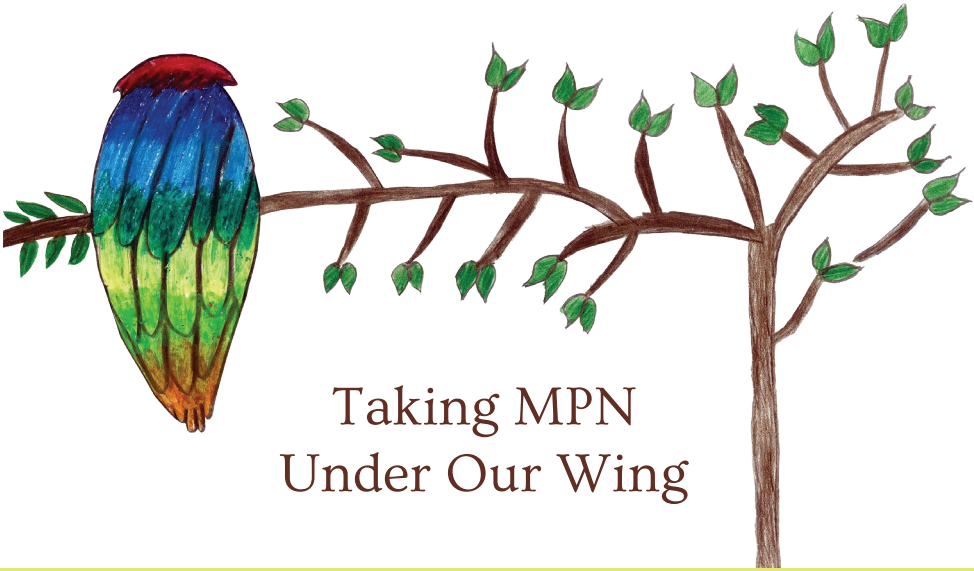
FOM Guide to Disease Management



FAQ on
Myelofibrosis

The Max Foundation

Friends of Max

FAQ

on

Myelofibrosis

Editor's Note

Dear Readers,

This FAQ on Myelofibrosis is the third in the series of Information Booklets on the various blood disorders that come under the classification of MPN.

Myelofibrosis is an MPN, a type of blood cancer which is characterized by the abnormal increase in bone marrow activity that disrupts the body's normal production of blood cells. There is significant scarring of bone marrow here which leads to low haemoglobin and platelets. Most patients develop pain or fullness below the ribs on the left side. This is due to an enlarged spleen. Excessive tiredness, night sweats and bone pain are other common manifestations of this condition. Some people with myelofibrosis have no symptoms and are diagnosed incidentally during evaluation for an unrelated cause. Myelofibrosis can affect anyone, but it's most often diagnosed in people older than 50 years. There is effective treatment for symptom control hence improving quality of life as well as life prolonging options like stem cell transplant leading to cure.

I am grateful to Friends of Max, for going out of their way in formulating this booklet. This will not only increase awareness of this illness but also resolve many queries of our patients.

- *Dr. Lingaraj Nayak,*
  Assistant Professor
  Department of Medical Oncology
  Tata Memorial Hospital, Mumbai

FAQ

What is Myelofibrosis (MF)?

Myelofibrosis is a rare type of blood cancer that disrupts the body's normal production of blood cells. This is characterized by the buildup of scar tissue, called "fibrosis," in the bone marrow. As scar tissue increases, the bone marrow is unable to make enough healthy blood cells. This leads to drop in hemoglobin which can cause weakness and fatigue. Bone marrow scarring can also lead to a low count of platelet which increases the risk of bleeding.

Broadly, Myelofibrosis is classified into primary and secondary. When Myelofibrosis develops on its own (and not as the result of another bone marrow disease) it is called Primary Myelofibrosis. When Myelofibrosis develops from a previous myeloproliferative disorder (MPN) like Polycythemia Vera (PV) or Essential Thrombocythemia (ET), then it is called Secondary Myelofibrosis.

Is Myelofibrosis a cancer?

Yes, Myelofibrosis is a type of cancer.

Is Myelofibrosis hereditary?

No, there is no evidence yet that Myelofibrosis is hereditary.

Can Myelofibrosis be passed on to another person in close regular contact?

No, it is not a contagious disease. For that matter, no cancer is contagious.

FAQ

Is Myelofibrosis curable?

Yes, is curable; the curative treatment being an allogeneic Stem Cell Transplant (SCT).

What are the causative/ risk factors of Myelofibrosis?

Myelofibrosis is a complex condition and researchers are still trying to discover its exact cause. However, we do know that there are certain abnormal proteins (called mutations) that can lead to myelofibrosis. Presence of these mutations help in diagnosis.

How is Myelofibrosis diagnosed?

Myelofibrosis is first considered a probable diagnosis after a thorough clinical examination and blood test. The definite diagnosis is done based on bone marrow studies. Bone marrow testing involves two steps usually performed simultaneously in a doctor's office or a hospital:

- A bone marrow aspiration to remove a liquid marrow sample
- A bone marrow biopsy to remove a small amount of bone filled with marrow

A pathologist studies the samples under the microscope and examines the chromosomes inside the cells. This is necessary to differentiate Myelofibrosis from other myeloproliferative disorders.

What are the stages of Myelofibrosis?

Myelofibrosis is a chronic, progressive disease. That means it doesn't go away, and it usually gets worse over time. In its early stages, Myelofibrosis may be silent. You may or may not experience symptoms even though the disease may be progressing. As the disease gets worse, however, the symptoms may also start to get worse.

FAQ

Who is affected by Myelofibrosis?

Although Myelofibrosis can occur in persons of any age, it is more common later in life. People are usually around 50 or older when they realise they have Myelofibrosis.

How common is Myelofibrosis?

Myelofibrosis is an uncommon cancer.

What is the life expectancy of Myelofibrosis patients?

The median life expectancy of patients diagnosed with Myelofibrosis is six years.

What can a patient newly diagnosed with Myelofibrosis expect?

Myelofibrosis affects each person differently. Some people have no symptoms. In others, Myelofibrosis symptoms can be severe.

Common symptoms are:

- Fatigue
- Early satiety (means unable to eat a full meal, or you feel very full after eating only a small amount of food)
- Abdominal discomfort
- Problems with concentration (as compared to prior routine)
- Intense sweating at night
- Itching (pruritus)
- Bone pain (diffuse- not to be confused with joint pain or arthritis)
- Fever (>100°F) that is persistent for beyond 4 weeks
- Unintentional or unexplained weight loss over six months

- Unexplained blood clots
- Abnormal bruising and bleeding

Most of the patients will have enlargement of spleen which can lead to following symptoms

- Abdominal discomfort in left upper side of tummy
- Pain under the left ribs
- Early feeling of fullness after eating a small amount of food

What are the common health complications due to Myelofibrosis?

- **Extramedullary haematopoiesis** — Formation of blood cells outside the bone marrow, called "extramedullary haematopoiesis," may create clumps (tumours) of developing blood cells in other areas of the body. These tumours may cause such problems as bleeding in stools with abdominal pain, coughing or spitting up blood, compression of vital organs like spinal cord.
- **Acute Myeloid Leukemia (AML)** — In a small fraction of patients with myelofibrosis, Myelofibrosis will change gears from a slow growing disease to AML (Acute myeloid leukemia), a type of blood and bone marrow cancer that progresses rapidly
- **Portal Hypertension** — Myelofibrosis commonly causes spleen enlargement. Sometimes, this spleen can become so big that it can lead to high blood pressure inside the portal vein, (it's a connection between spleen and liver). This increased pressure in portal vein can force excess blood into smaller veins in the stomach and oesophagus (food pipe), potentially causing the veins to rupture and bleed. This can lead to blood in vomitus, which may be life threatening.

FAQ

What are the treatment options available in India?

- Ruxolitinib
- Hydroxyurea
- Thalidomide
- Steroid medicines (like Danazol or others)

How is Myelofibrosis monitored?

Myelofibrosis is monitored based on the following:

1. Improvement in symptoms
2. Reduction in spleen size
3. Improvement in blood counts (commonly haemoglobin and platelets)

What are the common symptoms/warning signs of Myelofibrosis?

1. Rapid increase in spleen size leading to abdominal discomfort
2. Rapid drop in platelet counts or haemoglobin

Is it possible for one MPN to transform into another?

Yes. MPNs like Polycythaemia Vera (PV) and Essential Thrombocytosis (ET) can transform to Myelofibrosis.

What are the dietary restrictions for a Myelofibrosis patient?

There's no specific myelofibrosis diet. But eating a balanced diet that's rich in fruits, vegetables, whole grains, protein sources, and healthy fats may help lower inflammation and help people with myelofibrosis have more energy.

According to MPN Research Foundation, myelofibrosis patients must avoid over-consumption of processed and refined foods such as fast food, boxed sweets and sugared beverages.

What is the emotional impact of being diagnosed with Myelofibrosis and how can one seek help?

In addition to physical symptoms, MPNs affect emotional well-being, with anxiety and depression frequently reported by patients. One of the main issues impacting MPN treatment is the discord between patient and physician perceptions of symptom burden, treatment goals, and expectations. Additionally, regional variation further affects the psychosocial burden of MPNs on patients. Overcoming some of the challenges in patient–physician communication and treatment access are key to improving disease management and quality of life, as well as giving the patient greater input in treatment decisions.

Is it safe for a Myelofibrosis patient to take a vaccine/other medication?

It is generally safe to take vaccines. However, if a patient is on medicines like Ruxolitinib , they need to consult their treating oncologist/haematologist before taking any new drug or vaccine (to be aware of possible drug interaction.

Are there any physical restrictions that someone on treatment for Myelofibrosis must adhere to?

No, there are no physical restrictions for a patient on treatment for Myelofibrosis.

Acknowledgement

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Notes

Together we share & learn

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