

## ***Myelofibrosis***

Myelofibrosis (MF) is a chronic blood malignancy caused by abnormal blood cells built up in the bone marrow. Increased production of abnormal blood stem cells creates overproduction of megakaryocytes, which make platelets and fibroblast growth factor. Ultimately this causes the formation of fibrous tissue in the bone marrow. The fibrous tissue decreases the bone marrow's ability to make normal blood cells. Often, blood cell production then moves to other areas of the body such as the spleen and liver. Most patients exhibit symptoms of chronic inflammation due to "cytokines" released from the abnormal cells. MF is one of a group of rare blood cancers called myeloproliferative neoplasms (MPNs) that develop due to a stem cell mutation in the bone marrow.

### **Statistics**

- A majority of MF cases are diagnosed in people over the age of 60.
- About half of MF patients have a mutation in the JAK2 (Janus kinase) gene (V617F).
- About a quarter of patients have a Calreticulin (CALR) mutation.
- A MPL gene mutation is present in 5 to 10 percent of MF patients.

### **Risk Factors**

The exact causes of the acquired genetic mutation seen in MF are not yet known.

- MF can develop from other myeloproliferative diseases such as polycythemia vera (PV) or essential thrombocythemia (ET) and is then designated secondary or "post" PV or ET MF.
- Researchers are exploring the link between myelofibrosis and various gene mutations, such as JAK2, CALR, and MPL.
- Exposure to petrochemicals and radiation may increase risk for developing myelofibrosis.
- Primary MF carries a worse prognosis than secondary MF.

### **Symptoms**

Some patients with MF do not have symptoms when they are diagnosed, especially when the disease is in early stages. Diagnosis in an early phase often occurs during a routine exam or blood test. Many MF patients will have the following symptoms.

- Fatigue, weakness, or shortness of breath
- Anemia
- Unexplained weight loss
- Night sweats
- Bruising or easy bleeding
- Enlarged liver
- Frequent infections
- Discomfort in upper left side of abdomen, caused by an enlarged spleen and an abnormally quick feeling of fullness at meals
- Bone, joint pain, or gout
- Fever and itching
- Portal hypertension, which may cause enlarged veins in the stomach and esophagus and bleeding from the bowel
- Growth of blood-forming cells outside the bone marrow
- Teardrop red cells in the blood
- Early blood cell precursors (nucleated red blood cells and mature white blood cells, normally only in the bone marrow) found in the blood

### **Prevention**

MF cannot be prevented. Research is underway to learn more about how the disease develops.

### **Treatment**

MF patients who do not experience symptoms typically do not receive treatment, and may remain in stable health for years. However, if symptoms are present, treatment options include hydroxyurea, ruxolitinib, interferon, blood transfusions, and less often, radiation, splenectomy, and stem cell transplants. Clinical trials can also be an important treatment option.

*Sources: Leukemia and Lymphoma Society, MPN Research Foundation, and National Cancer Institute*



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