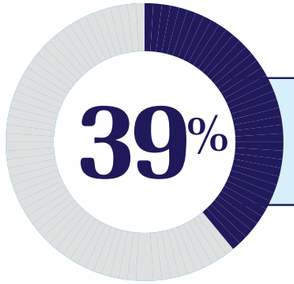
# Start Implementing a Quality Initiative for Patients with Myelofibrosis (MF)

## A GUIDE FOR PHARMACY DIRECTORS AND CLINICAL PHARMACISTS

Proactively identify and support patients with symptoms associated with MF in need of better management

# Myelofibrosis (MF) is a serious hematologic malignancy

MF is a Philadelphia chromosome-negative myeloproliferative neoplasm (MPN) marked by bone marrow fibrosis, abnormal blood counts, extramedullary hematopoiesis, a significant symptom burden, and shortened survival.[1]




## 5-YEAR OVERALL SURVIVAL (OS)

**39% of patients with MF reach 5-year OS[2,a]**

# Most patients with MF have intermediate or high-risk disease, which is associated with shortened survival[3]

**Any one of the following risk factors[b] indicates the patient is already at intermediate risk[3]:**

- Hemoglobin level <10 g/dL
- Circulating blast cells ≥1%
- Leukocyte count >25 x $10^9$/L
- Platelet count <100 x $10^9$/L
- Age >65 years
- Constitutional symptoms
- Red cell transfusion dependency
- Unfavorable karyotype

## INTERMEDIATE OR HIGH-RISK AT DIAGNOSIS

**95%** **of 491 patients diagnosed with MF** in a retrospective chart review sponsored by Incyte **were at intermediate or high risk at diagnosis[4]**

**90%** **of 428 evaluable patients with primary MF**, in a separate study, **were considered to be at intermediate or high risk *within 1 year of diagnosis*[3]**

Hb, hemoglobin; MPN-SAF TSS, Myeloproliferative Neoplasm-Symptom Assessment Form Total Symptom Score; NCCN, National Comprehensive Cancer Network; PLT, platelet.

[a] 5-year overall survival rate was estimated using Surveillance, Epidemiology, and End Results (SEER) data obtained from population-based cancer registries of the US population and SEER*Stat Software version 8.3.2. The analysis included patients with initial/primary site diagnosis between years 2007-2011. Overall survival is defined as the proportion of patients surviving at the specified time interval after diagnosis.[2]

[b] As included in the Dynamic International Prognostic Scoring System (DIPSS) Plus tool. The DIPSS-Plus scoring system has been validated for risk stratification any time after a diagnosis of primary MF, but has been used clinically for risk stratification of patients with post-essential thrombocythemia MF and post-polycythemia vera MF. In the DIPSS-Plus scoring system, adverse points are assigned by first calculating the DIPSS score and then adding points for additional factors.

To calculate the DIPSS score, 1 point each is assigned to age >65 years, leukocyte count >25 × $10^9$/L, circulating blast cells ≥1%, and constitutional symptoms (weight loss greater than 10% of the baseline value in the year preceding the primary MF diagnosis and/or unexplained persistent fever or excessive sweating), while 2 points are assigned for anemia (Hb <10 g/dL).

A DIPSS risk category is calculated, where 0 points = low risk, 1 or 2 points = intermediate-1 risk, 3 or 4 points = intermediate-2 risk, and 5 or 6 points = high risk. The DIPSS risk categories—low, intermediate-1, intermediate-2, and high risk—are given 0, 1, 2, or 3 points, respectively, in the DIPSS-Plus system, with an additional 1 point each for PLT count <100 × $10^9$/L, red cell transfusion dependency, or unfavorable karyotype (complex karyotype or single or 2 abnormalities including +8, –7/7q-, i(17q), –5/5q-, 12p-, inv(3) or 11q23 rearrangement), resulting in a maximum possible score of 6.[3]

# Majority of patients with MF report symptom burden at diagnosis[5,6]

## PREVALENCE OF SYMPTOMS AT DIAGNOSIS

**95%** **of patients reported 2+ MF-related symptoms at diagnosis**

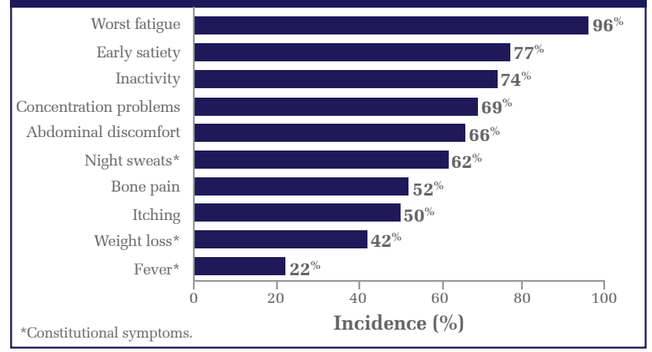
*based on a retrospective chart review of 180 patients with MF[5*]*

*Retrospective, observational study of symptom burden and splenomegaly in 180 patients with MF; data were collected at the time of diagnosis of MF in patients without splenomegaly (n=78) or at the time of detection of splenomegaly in patients with splenomegaly (n=102). In patients with splenomegaly, splenomegaly was most often recorded at the time of diagnosis (median time from MF diagnosis to reported splenomegaly was 1 day).[5]

## Burden of symptoms in MF

- In the MPN Landmark survey, many patients with MF (49%) reported experiencing symptoms at least 1 year before diagnosis[6,c]
- Symptoms may be present even in patients with earlier disease[5,6]

### SELF-REPORTED SYMPTOMS OF MF[7,d]




*Constitutional symptoms.

Patient-reported results from the MPN Landmark Survey[6]:

## THE MAJORITY OF PATIENTS WITH MF REPORTED THAT SYMPTOMS IMPACT QUALITY OF LIFE[6]

**81% reported that their symptoms reduced their quality of life[6]**

**79% reported that MF interfered with family or social life[6,e]**

[c] The MPN Landmark Survey, funded by Incyte Corporation, was a web-based questionnaire composed of 65 multiple-choice questions intended to help evaluate the patient's perception of disease burden in the MPN disease setting. A total of 813 patients in the United States with a previous diagnosis of polycythemia vera (n = 380), MF (n = 207), or essential thrombocythemia (n = 226) participated.[6]

[d] This prospective study included a total of 1433 patients with MPNs (n = 293 with MF), who were queried on the 10 symptoms from the MPN-SAF TSS/MPN-10. The MPN-SAF TSS is validated for serial tracking of the most pertinent MPN-related symptoms—fatigue, concentration problems, early satiety, inactivity, night sweats, itching, bone pain, abdominal discomfort, weight loss, and fever—scored on a scale of 0 (absent/as good as it can be) to 10 (worst imaginable/as bad as it can be), for a total possible score of 100.[7]

[e] Patients reported impact on their activities of daily living on a scale that ranged from 1 (not at all) to 5 (a great deal).[6]

# Assessing symptoms in MF

**NCCN Clinical Practice Guidelines in Oncology (NCCN GUIDELINES®)** recommend assessing symptoms (in a provider's office) at baseline and monitoring symptom status (stable, improved, or worsening) during the course of treatment.[8]

**Changes in symptom status could be a sign of disease progression.[7]**

# Patients may not recognize that their symptoms are related to MF.[9] Quality Initiatives can help.[4,10]

Use this established sample workflow to proactively monitor patients with MF for symptom burden[10]:


A large regional health facility used this approach to proactively identify and better manage patients with MF by looking for those whose symptoms were unrecognized. A partnership between physicians and specialty pharmacists is feasible and can be successful. A multidisciplinary approach incorporating telemedicine for MF patients provides an effective method to measure patient symptom burdens and to assign prognostic categories.[10]

**How can you apply these learnings to implement a Quality Initiative in patients with MF in your practice today?**

**Visit MPNQuality.com today to see videos and download information on the importance of implementing Quality Initiatives in MF**

SCAN ME

EMR, electronic medical record.

**References: 1.** Vannucchi AM, Guglielmelli P, Tefferi A. Advances in understanding and management of myeloproliferative neoplasms. *CA Cancer J Clin*. 2009;59(3): 171-191. doi:10.3322/caac.20009 **2.** Surveillance, Epidemiology, and End Results (SEER) Program. SEER*Stat database: incidence - SEER 18 Regs research data + Hurricane Katrina impacted Louisiana cases, November 2015 submission (1973-2013 varying) - linked to county attributes - total US, 1969-2014 counties. National Cancer Institute. Accessed March 15, 2023. http://www.seer.cancer.gov **3.** Gangat N, Caramazza D, Vaidya R, et al. DIPSS plus: a refined Dynamic International Prognostic Scoring System for primary myelofibrosis that incorporates prognostic information from karyotype, platelet count, and transfusion status. *J Clin Oncol*. 2011;29(4):392-397. doi:10.1200/JCO.2010.32.2446 **4.** Verstovsek S, Yu J, Kish JK, et al. Real-world risk assessment and treatment initiation among patients with myelofibrosis at community oncology practices in the United States. *Ann Hematol*. 2020;99(11):2555-2564. doi:10.1007/s00277-020-04055-w **5.** Mitra D, Kaye JA, Piecoro LT, et al. Symptom burden and splenomegaly in patients with myelofibrosis in the United States: a retrospective medical record review. *Cancer Med*. 2013;2(6):889-898. doi:10.1002/cam4.136 **6.** Mesa R, Miller CB, Thyne M, et al. Myeloproliferative neoplasms (MPNs) have a significant impact on patients' overall health and productivity: the MPN Landmark survey. *BMC Cancer*. 2016;16:167. doi:10.1186/s12885-016-2208-2 **7.** Emanuel RM, Dueck AC, Geyer HL, et al. Myeloproliferative neoplasm (MPN) symptom assessment form total symptom score: prospective international assessment of an abbreviated symptom burden scoring system among patients with MPNs. *J Clin Oncol*. 2012;30(33):4098-4103. doi:10.1200/JCO.2012.42.3863 **8.** Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Myeloproliferative Neoplasms V.3.2022. © National Comprehensive Cancer Network, Inc 2022. All rights reserved. Accessed January 5, 2023. To view the most recent and complete version of the guideline, go online to NCCN.org. NCCN makes no warranties of any kind whatsoever regarding their content, use or application and disclaims any responsibility for their application or use in any way. **9.** Mesa RA, Miller CB, Thyne M, et al. Differences in treatment goals and perception of symptom burden between patients with myeloproliferative neoplasms (MPNs) and hematologists/oncologists in the United States: findings from the MPN Landmark survey. *Cancer*. 2017;123(3):449-458. doi:10.1002/cncr.30325 **10.** Chojecki AL, DiSogra KY, Arnall J, et al. Optimization of physician and specialty pharmacy clinical workflow in assessment of risk category and symptom burden in patients with myelofibrosis (MF). *Leuk Lymphoma*. 2022;63(6):1515-1517. doi:10.1080/10428194.2022.2032037.

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