
بسم الله الرحمن الرحیم

Allogenic HSCT In High Risk Myeloproliferative Disorders

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Higher-risk MF

- Patients with higher-risk MF have a poor prognosis and are likely to have significant disease-associated symptoms.
- Management is guided by eligibility for allogeneic hematopoietic cell transplantation (HCT).
- Many institutions limit allogeneic HCT to patients ≤70 years without major medical comorbidities.

ve **NCCN Guidelines Version 1.2025**
Myelofibrosis

RISK STRATIFICATION FOR PATIENTS WITH MYELOFIBROSIS

PMF

DIPSS	(MF-A, 2 of 5)
DIPSS-PLUS	(MF-A, 2 of 5)
MIPSS-70	(MF-A, 3 of 5)
MIPSS-70+ Version 2.0	(MF-A, 4 of 5)

POST-PV AND POST-ET MF

MYSEC-PM	(MF-A, 5 of 5)

MIPSS70+v2.0-Mutation-enhanced International Prognostic Scoring System plus karyotype ,version2.0

1. **Karyotype**
   - VHR karyotype (4 points)
   - Unfavorable karyotype (3 points)
   - Other karyotype (0 points)
2. **HMR mutations**
   - â‰¥2 HMR mutations (3 points)
   - 1 HMR mutation (2 points)
   - No HMR mutations (0 points)
3. **Absence of type 1/like CALR mutation**
   - Yes (2 points)
   - No (0 points)
4. **Presence of constitutional symptoms**
   - Yes (2 points)
   - No (0 points)
5. **Anemia severity**
   - Severe anemia (2 points)
   - Moderate anemia (1 point)
   - Mild or no anemia (0 points)
6. **>2% circulating blasts**
   - Yes (1 point)
   - No (0 points)

MIPSS70+v2.0

MIPSS70+ version 2.0 score (points)	Risk group	10-year overall survival	Median overall survival (years)
0	Very low risk	86%	Not reached
1 to 2	Low risk	50%	10.3
3 to 4	Intermediate risk	30%	7
5 to 8	High risk	10%	3.5
9 to 14	Very high risk	<3%	1.8

MIPSS70+v2.0 : Note

Cytogenetics:

1. **Very high risk (VHR):** Single/multiple abnormalities of -7, i(17q), inv(3)/3q21, 12p-/12p11.2, 11q-/11q23, or other autosomal trisomies not including+8/+9 (eg, +21, +19).
2. **Favorable:** Normal karyotype or sole abnormalities of 13q-, +9, 20q-, chromosome 1 translocation/duplication or sex chromosome abnormality including -Y.
3. **Unfavorable:** All other abnormalities.

High molecular risk (HMR) mutations: *ASXL1, SRSF2, EZH2, IDH1, IDH2, U2AF1* Q157.

GIPSS: Genetically Inspired Prognostic Scoring System

- **Karyotype classification:**
  - Very high risk (VHR) (2 points)
  - Unfavorable (1 point)
  - Favorable (0 points)
- **Driver mutations:**
  - Absence of type 1-like CALR (1 point)
- **High molecular risk (HMR) mutations:**
  - ASXL1 mutation (1 point)
  - SRSF2 mutation (1 point)
  - U2AF1 Q157 mutation (1 point)

GIPSS score (points)	Risk group	5-year overall survival	Median overall survival (years)
0	Low risk	94%	26.4
1	Intermediate risk - 1	73%	10.3
2	Intermediate risk - 2	40%	4.6
3 to 6	High risk	14%	2.6

- Use of other prognostic models, such as DIPSS (Dynamic International Prognostic Scoring System) or DIPSS-Plus , is acceptable where neither cytogenetics nor molecular analysis is available.

DIPSS And DIPSS Plus For Primary Myelofibrosis

DIPSS score

The DIPSS score assigns points for the following five variables:

- Age >65 years: 1 point
- Leukocyte count >25,000/microL (>25 x 10^9/L): 1 point
- Hemoglobin <10 g/dL (<100 g/L): 2 points
- Circulating blast cells â‰¥1%: 1 point
- Constitutional symptoms*: 1 point

The resulting score is interpreted as follows:

- 0 points â€" low risk
- 1 to 2 points â€" intermediate-1 risk
- 3 to 4 points â€" intermediate-2 risk
- 5 to 6 points â€" high risk

DIPSS Plus score

The DIPSS Plus score assigns points for the following variables

- DIPSS low risk: 0 points
- DIPSS intermediate-1 risk: 1 point
- DIPSS intermediate-2 risk: 2 points
- DIPSS high risk: 3 points
- Unfavorable karyotype[Â¶]: 1 point
- Platelet count <100,000/microL (<100 x 10^9/L): 1 point
- Anemia requiring transfusion: 1 point

The resulting score is interpreted as follows:

- 0 points â€" low risk
- 1 point â€" intermediate-1 risk
- 2 to 3 points â€" intermediate-2 risk
- 4 to 6 points â€" high risk

MYSEC Prognostic Model Risk Calculator (MYSEC-PM)

Age at diagnosis

Haemoglobin < 11 g/dL [+2 pt]

Platelets < 150 × 10^9/L [+1 pt]

Blast>3% [+2 pt]

CALR -unmutated genotype [+2 pt]

Constittional symptom[+1 pt]

Prognostic Classification

Higher-risk PMF :

- **GIPSS**
  - High risk
  - Intermediate (int)-2
- **MIPSS70+ v2.0**
  - Very high risk
  - High risk

Lower-risk PMF :

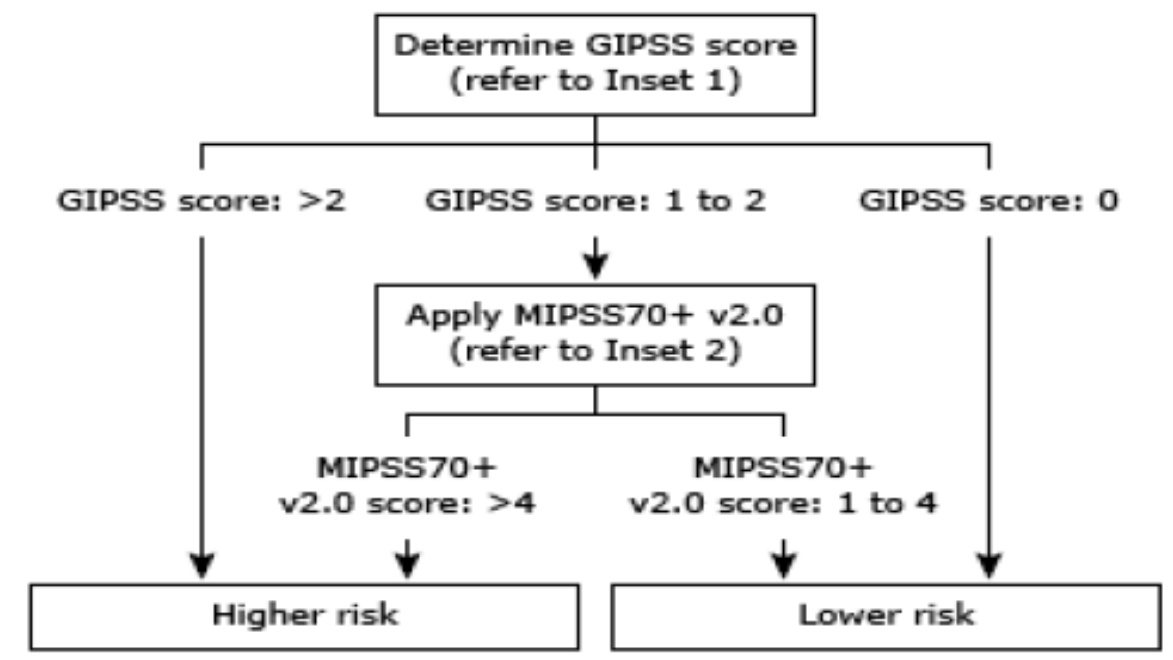
- **GIPSS**
  - Low risk
  - Int-1
- **MIPSS70+ v2.0**
  - Intermediate risk
  - Low risk
  - Very low risk

Risk Stratification In Primary Myelofibrosis

Inset 1

GIPSS score (sum of points below):

- Karyotype:*
  - Very high risk (2 points)
  - Unfavorable (1 point)
- Driver mutations:
  - Absence of type 1-like *CALR* (1 point)
- HMR mutations:
  - *ASXL* mutation (1 point)
  - *SRSF2* mutation (1 point)
  - *U2AF1* Q157 mutation (1 point)

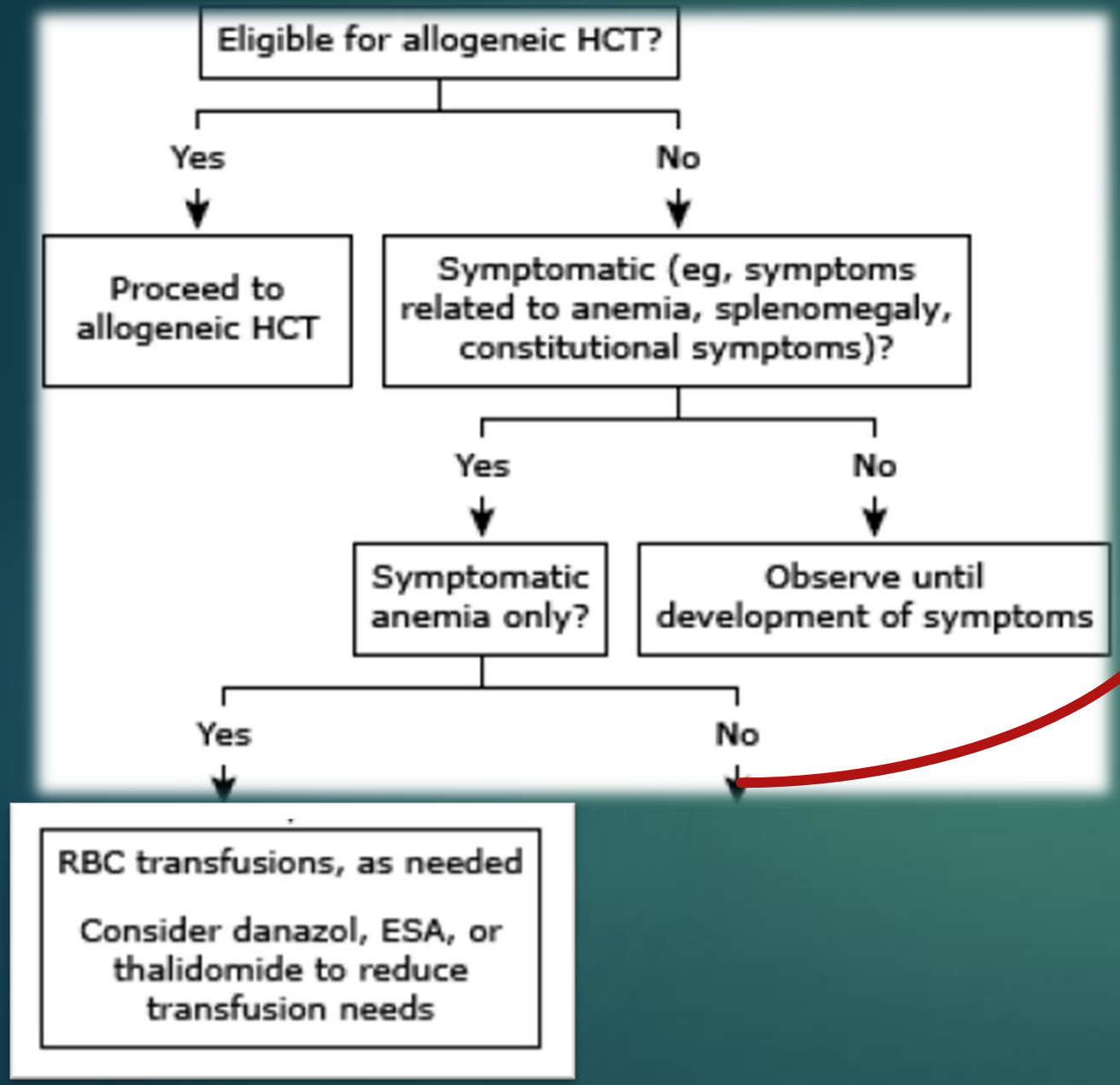


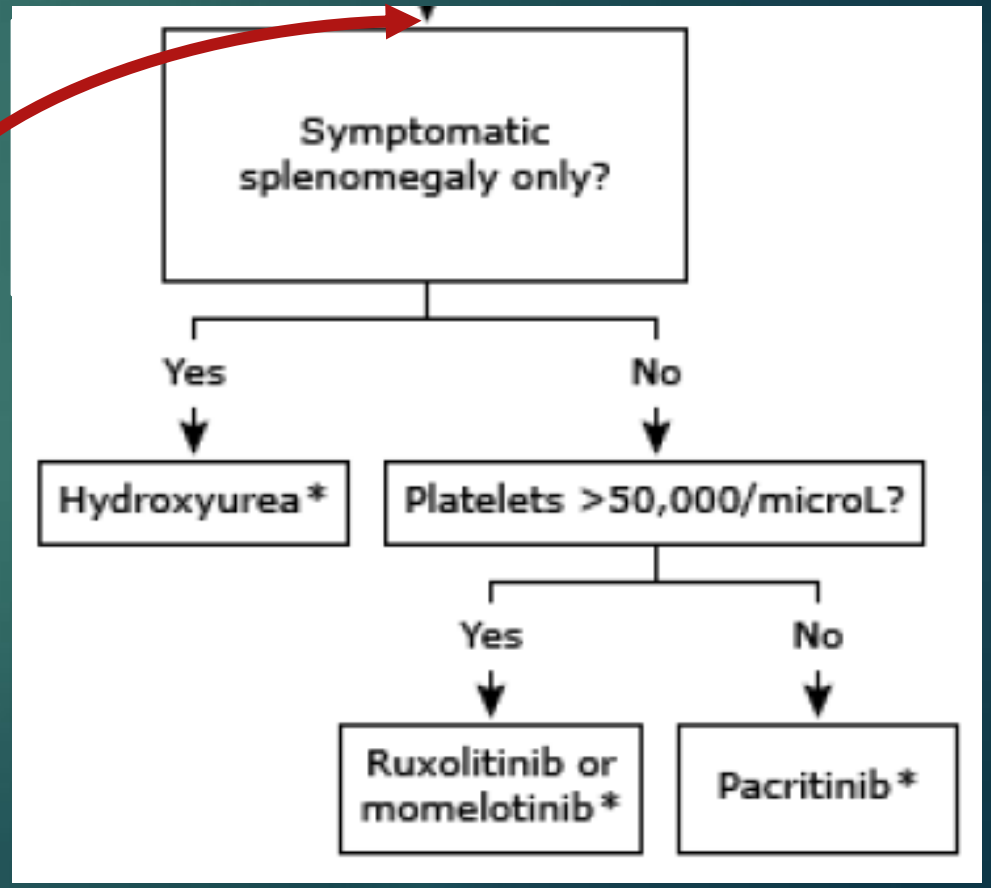
Inset 2

MIPSS70+ v2.0 score (sum of points below):

- Clinical:
  - Severe anemia: Men Hb <9 g/dL, women Hb <8 g/dL (2 points)
  - Moderate anemia: Men Hb 9 to 10.9 g/dL, women Hb 8 to 9.9 g/dL (1 point)
  - Circulating blasts ≥2% (1 point)
  - Constitutional symptoms (2 points)
- Karyotype:
  - Very high risk (4 points)
  - Unfavorable (3 points)
- Mutations:
  - >1 HMR mutations (3 points)
  - 1 HMR mutation (2 point)
  - Absence of type 1-like *CALR* mutation (2 points)

Primary and secondary myelofibrosis: Management of higher-risk disease





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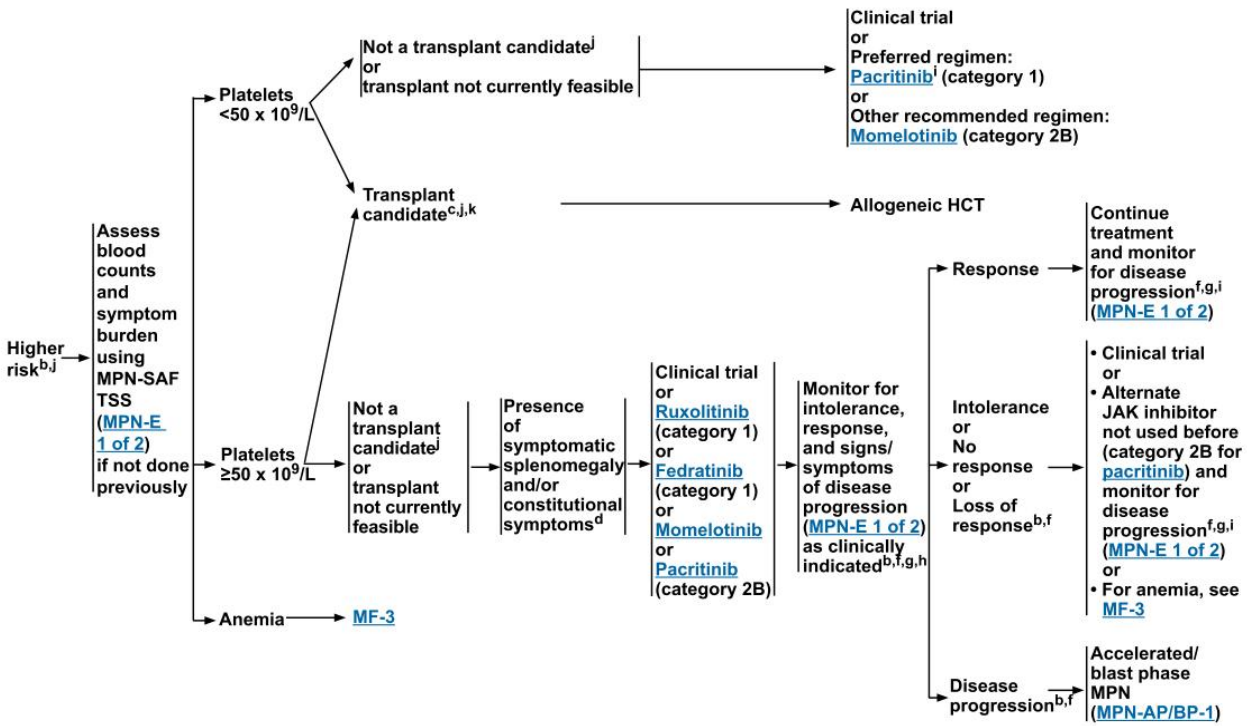

Note: All recommendations are category 2A unless otherwise indicated.

Footnotes on MF-2A

Thanks For Your Attention