

TABLE

Contemporary prognostic scoring systems for primary myelofibrosis

Models	Variables	Median Survival by Risk Category					
		Very Low	Low	Intermediate-1	Intermediate-2	High	Very High
IPSS^d <i>International Prognostic Scoring System</i>	Age >65 years (1 point) Constitutional symptoms ^a (1 point) Hemoglobin <10 g/dL (1 point) Leukocytes >25 × 10 ⁹ /L (1 point) Circulating blasts ≥1% (1 point)	NA	(0 points) 11.3 years	(1 point) 7.9 years	(2 points) 4 years	(≥3 points) 2.3 years	NA
DIPSS^e <i>Dynamic International Prognostic Scoring System</i>	Age >65 years (1 point) Constitutional symptoms (1 point) Hemoglobin <10 g/dL (2 points) Leukocytes >25 × 10 ⁹ /L (1 point) Circulating blasts ≥1% (1 point)	NA	(0 points) 15.4 years	(1 point) 6.5 years	(2–3 points) 2.9 years	(≥4 points) 1.3 years	NA

Models	Variables	Very Low	Low	Intermediate-1	Intermediate-2	High	Very High
DIPSS-plus^e	Age > 65 years (1 point) Constitutional symptoms ^a (1 point) Hemoglobin <10 g/dL (1 point) Leukocytes >25 × 10 ⁹ /L (1 point) Circulating blasts ≥1% (1 point) Unfavorable karyotype ^h (1 point) Platelet count <100 × 10 ⁹ /L (1 point) Transfusion needs (1 point)	NA	(0 points) 15.4 years	(1 point) 6.5 years	(2-3 points) 2.9 years	(≥4 points) 1.3 years	NA
MIPSS70^d <i>Mutation-enhanced International Prognostic Scoring System (Age ≤ 70 years)</i>	≥2 HMR mutations ^b (2 points) Leukocytes >25 × 10 ⁹ /L (2 points) Platelets <100 × 10 ⁹ /L (2 points) Hemoglobin <10 g/dL (1 point) Circulating blasts ≥2% (1 point)	NA	(0–1 point) Not reached	(2–4 points) 6.3 years		(≥5 points) 3.1 years	NA

Models	Variables	Very Low	Low	Intermediate-1	Intermediate-2	High	Very High
MIPSS70^d <i>Cont.</i>	BM fibrosis grade ≥ 2 (1 point) Constitutional symptoms ^a (1 point) Type 1/like <i>CALR</i> absent (1 point) One HMR mutation ^b (1 point)						
MIPSS70+ v2^e	Very high-risk karyotype ^f (4 points) Unfavorable karyotype ^g (3 points) ≥ 2 HMR mutations ^c (3 points) One HMR mutation ^c (2 points) Type 1/like <i>CALR</i> absent (2 points) Constitutional symptoms ^a (2 points) Severe anemia ⁱ (2 points) Moderate anemia ^j (1 point) Circulating blasts $\geq 2\%$ (1 point)	(0 points) Not reached	(0 points) 16.4 years	(3-4 points) 7.7 years		(5-8 points) 4.1 years	(> 9 points) 1.8 years

Models	Variables	Very Low	Low	Intermediate-1	Intermediate-2	High	Very High
GIPSS^e <i>Genetics-inspired International Prognostic Scoring System</i>	Very high-risk karyotype ^f (2 points) Unfavorable karyotype ^g (1 point) ASXL1 mutation (1 point) SRSF2 mutation (1 point) U2AF1Q157 mutation (1 point) Type 1/like CALR absent (1 point)	NA	(0 points) 26.4 years	(1 point) 8 years	(2 points) 4.2 years	(≥3 points) 2 years	NA

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^a **Constitutional symptoms:** weight loss, fever, drenching night sweats.

^b **High molecular risk (HMR) mutations** for MIPSS70 include ASXL1, SRSF2, EZH2, IDH1, IDH2.

^c **HMR for MIPSSv2 and GIPSS** include ASXL1, SRSF2 and U2AF1Q157.

^d Parameters used at time of diagnosis.

^e Parameters used at any time in the clinical course.

^f **Very high-risk (VHR) karyotype** = single/multiple abnormalities of -7, inv (3)/3q21, i(17q), 12p-/12p11.2 or 11q-/11q23, single/multiple autosomal trisomies other than +9 and +8.

^g **Unfavorable karyotype in the context of GIPSS/MIPSSv2** = any abnormal karyotype other than VHR karyotype, normal karyotype or sole abnormalities of 20q-, 13q-, +9, chr. 1 translocation/duplication, -Y, or sex chromosome abnormality other than -Y.

^h **Unfavorable karyotype in the context of DIPSS-plus** = complex karyotype or sole or two abnormalities that include +8, -7/7q-, i(17q), inv (3), -5/5q-, 12p- or 11q23 rearrangement any.

ⁱ **Severe anemia:** Hemoglobin <8 g/dL in women and <9 g/dL men.

^j **Moderate anemia:** Hemoglobin 8–9.9 g/dL in women and 9–10.9 g/dL men.