**Epo/G-CSF in low/int-I risk MDS \*\* Pre-treatment evaluation**

Indicatie: Hb <6,2 en score 0-1

transfusion need ≥ 2 U/month: 1 pt

serum-Epo ≥ 200 U/liter: 1 pt (of 500?)

predicted response rate: total score 0=74%, 1=23%, 2=7%

Epoëtine bèta (Epo; NeoRecormon) sc 30000 IU weekly for 3 months

after 6 weeks dose escalation of 30000 IU if Hb not +0.6 mmol/l

after 12 weeks, G-CSF (filgrastim; Neupogen) in combination with Epo

G-CSF 3 times weekly (3x300 μg/week sc <75kg; 3x480 μg/week sc >75 kg)` G-CSF interrupted if WBC >30x109/l, restart after normalization of WBC

G-CSF re-started at 300 μg once a week, escalated to 2x300 μg/wk in 6 wks

complete remission (Hb > 6,8 o.a.)

Epo reduced with 10000 IU/week

each reduction lasting for 6 weeks

minimum maintenance dose of 20000 IU/week

**WHO 2008**



**International Prognostic Scoring System (IPSS)**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Prognostische variabele** | **score** |   |   |   |   |
|   | **0** | **0.5** | **1.0** | **1.5** | **2.0** |
| BM blasten (%) | < 5 | 5-10  | -  | 11-20  | 21-30 |
| Karyotype\* |  Goed |  Intermediar |  Slecht |   |   |
| Cytopenieën |  0/1 |  2/3 |   |   |   |

|  |  |  |
| --- | --- | --- |
| **IPSS groep** | **Score** | **Levensverwachting (jr med)** |
| **Laag** | 0 | 5.7 |
| **Int-1** | 0.5-1.0 | 3.5  |
| **Int-2** | 1.5-2.0 | 1.2 |
| **Hoog** | >2.0 | 0.4 |

**WPSS en WPSS-refined / Malcovati**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|   | **Score** |   |   |   |
| **Variabele** | **0** | **1** | **2** | **3** |
| WHO 2001  |  RA, RARS,del(5q) |  RCMD+/-RS |  RAEB-1 |  RAEB-2 |
| Karyotype |  goed |  intermed |  slecht |   |
| Transfusie / Hb | Nee of Hb> grenswaarde | Ja of Hb< grenswaarde |   |   |

**Extra punten:** extra 1 scorepunt bij matige/ernstige beenmergfibrose en/of comorbiditeit

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Risicogroep** | Zeer laag | Laag   | Intermed | Hoog | Zeer hoog |
| Score |  0 | 1 | 2 | 3-4 | 5-6 |



**Revised international prognostic scoring system (IPSS-R) in myelodysplastic syndrome**

|  |  |
| --- | --- |
| **Prognostic variable** | **Score** |
| **0** | **0.5** | **1.0** | **1.5** | **2.0** | **3.0** | **4.0** |
| Cytogenetics\* | Very good |  | Good |  | Intermediate | Poor | Very poor |
| Bone marrow blast (percent) | ≤2  |  | >2 to <5  |  | 5 to 10  | >10  |  |
| Hemoglobin (g/dL) | ≥10 |  | 8 to <10 | <8 |  |  |  |
| Platelets (cells/microL) | ≥100 | 50 to 100 | <50 |  |  |  |  |
| Absolute neutrophil count (cells/microL) | ≥0.8 | <0.8 |  |  |  |  |  |
| This scoring system was applied to an initial group of 7012 patients with primary MDS by the French-American-British classification who had at least two months of stable blood counts, ≤30 percent bone marrow blasts and ≤19 percent peripheral blood blasts, and who were observed until progression to AML transformation or death (did not receive disease-modifying agents for MDS). Patients could be stratified into five groups with the following estimated overall survival and progression to AML. |
| **Risk group** | **IPSS-R score** | **Median overall survival (years)** | **Median time to 25 percent AML evolution (years)** |
| Very low | ≤1.5 | 8.8 | >14.5 |
| Low | >1.5 to 3.0 | 5.3 | 10.8 |
| Intermediate | >3 to 4.5 | 3.0 | 3.2 |
| High | >4.5 to 6 | 1.6 | 1.4 |
| Very high | >6 | 0.8 | 0.7 |
|  |

It has been concluded that young patients with two or more adverse features (ie, hemoglobin <10 g/dL, constitutional symptoms, isolated cytogenetic abnormality, or blasts >1 percent) [[9,52](http://www.uptodate.com/contents/prognosis-and-treatment-of-primary-myelofibrosis/abstract/9%2C52)] should be considered for HCT shortly after diagnosis.