**MDS Prognostic Scoring System: IPSS-R**

<table>
<thead>
<tr>
<th>Prognostic Variables</th>
<th>Lille</th>
<th>IPSS</th>
<th>DIPSS</th>
<th>DIPSS-Plus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anemia (Hgb &lt; 10g/dl)</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td>x</td>
</tr>
<tr>
<td>WBC &gt; 25 x 10^9/L</td>
<td>x*</td>
<td>x</td>
<td>x</td>
<td>x</td>
</tr>
<tr>
<td>Circulating blasts ≥ 1%</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Constitutional symptoms</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Age &gt; 65</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Abnormal karyotype</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Platelets &lt;100 x 10^9/L</td>
<td>x</td>
<td></td>
<td></td>
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<tr>
<td>RBC transfusion dependence</td>
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</tbody>
</table>

**Prognosis in Primary Myelofibrosis**

- Anemia (Hgb < 10g/dl)
- WBC > 25 x 10^9/L
- Circulating blasts ≥ 1%
- Constitutional symptoms
- Age > 65
- Abnormal karyotype
- Platelets <100 x 10^9/L
- RBC transfusion dependence

*The Lille score identified a WBC > 30 x 10^9 or a WBC of < 4 x 10^9 as adverse indicators*

<table>
<thead>
<tr>
<th>Prognostic Variables in CMML</th>
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</thead>
<tbody>
<tr>
<td><strong>Hgb &lt; 12 g/dl</strong></td>
</tr>
<tr>
<td><em>Circulating immature myeloid cells</em></td>
</tr>
<tr>
<td><em>Absolute lymphocyte count &gt; 2.5 x 10^9/L</em></td>
</tr>
<tr>
<td>≥ 10% marrow blasts</td>
</tr>
<tr>
<td><strong>Cytogenetic risk category</strong></td>
</tr>
<tr>
<td>Low: Normal or −Y</td>
</tr>
<tr>
<td>High: +8, abn of 7, complex</td>
</tr>
<tr>
<td>Intermediate: All others</td>
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</tbody>
</table>

*Other prognostic scoring systems such as WPSS include WHO risk classification, transfusion dependence, and cytogenetic class. The MDACC model incorporates age, performance status, leukocytosis, degree of anemia/thrombocytopenia, marrow blasts, and cytogenetic class.*

MDS: Refractory Cytopenias and Dysplasia

Clinical features
- Constitutional symptoms less likely
- Refractory cytopenia(s)
- Organomegaly unusual

Blood and marrow findings
- Hyper > hypocellularity
- ↑Peripheral/marrow blasts
- Single or multi-lineage dysplasia
- +/- ring sideroblasts

Cytogenetic and molecular features
- *CKA in 40-70%, even in absence of dysplasia (commonly 5q-, -7 or 7q-, +8, 20q-, and -Y)
- JAK2 V617F infrequent (5%)

MDS/MPN Overlap Syndromes: Dysplastic Cytopenias with Proliferation

Clinical features
- +/- Constitutional symptoms cytopenia(s) with cytosis:
  - Thrombocytosis (RARS-T)
  - Monocytosis (CMML)
  - Neutrophilia (atypical CML)
  - Organomegaly

Blood and marrow findings
- Marrow hypercellularity
- ↑Peripheral/marrow blasts
- Dysplasia
- Ring sideroblasts (RARS-T)

Cytogenetic and molecular features
- CKA in ~30% of CMML
- (+8, del(5q), +10, -11q, -12p, +17p, +19, and +21, and -Y)
- JAK2 V617F mutation (~50%) in RARS-T

Myelofibrosis: Myeloproliferation

Clinical features
- Constitutional symptoms most likely
- Cytopenia or cytosis
- Increased LDH
- Organomegaly

Blood and marrow findings
- Leukoerythroblastosis
- ↑Peripheral/marrow blasts
- Variablecellularity
- Proliferating/atypical megakaryocytes
- Reticulin/collagen fibrosis

Cytogenetic and molecular features
- CKA in ~50%
- (20q-, 13q-, abn chromosome 1 or 12, +8, +9, -5, and -7)
- JAK2 V617F (60%) and MPL (<10%)