

Myelodysplastic Syndromes: Disease Snapshot

Feature	Key Findings		
Epidemiology	15,000-20,000 new cases each year with 35,000-50,000 existing cases Average age at diagnosis: 76 years		
Etiology	Genetic instability Chemical exposure Tobacco	Mutagens Autoimmune disease Unknown in majority of cases (~80%)	
Stem cell defect Myeloid progenitor cell	Intrinsic factors: Malignant clone Cytogenetic abnormalities Epigenetic DNA modification (hypermethylation)	Extrinsic factors: Bone marrow microenvironment Stromal dysregulation Cytokine abnormalities Imbalance of apoptosis and proliferation	
Chromosomal findings Cytogenetic abnormality present in ~ 40%	Favorable -Y, del(5q), -20q	Intermediate risk +8 and other	Poor risk Complex (> 3 abnormalities), chromosome 7 abnormalities: 7q, -7, del(7p), inv16, t(8:12)—AML
Additional prognostic factors indicating high-risk disease	<ul style="list-style-type: none"> • Increased transfusion burden (> 2 units in 4 weeks) • Increased blast cells (> 20% implies leukemic transformation) • Severe thrombocytopenia or neutropenia at diagnosis • CD7, CD117, CD56, CD44 expression by flow cytometry • Atypical localization of immature precursors (ALIP) • Ongoing analysis of more sensitive testing for chromosomal and molecular attributes 		
Staging	FAB/WHO (morphology) and IPSS/WPSS (risk stratification)		
Response criteria	International Working Group (IWG) criteria 2006		
Disease characteristics (all are incurable)	IPSS Low-Intermediate 1 risk Indolent course Low probability of leukemic transformation	IPSS Intermediate 2-High risk Progressive course with early transformation to acute leukemia	
Clinical presentation	Cytopenias— anemia most common Fatigue	Infection Bleeding	
Indication to treat	Transfusion dependence, progressive or symptomatic cytopenias, increased blasts		
Key concepts for effective treatment	<ul style="list-style-type: none"> • Supportive care alone does not prevent disease progression (no effect on underlying disease) • Active therapies for MDS generally require a minimum of 4-6 months to achieve response; premature discontinuation may limit potential for an optimal response • Aggressive concurrent management of cytopenias is essential to effective therapy • Treatment goals: reduce transfusion requirements, delay time to leukemic transformation, and improve quality of life; prolong survival • Chromosomal abnormalities have prognostic value 		
FDA-approved therapies	Azacitidine	Decitabine	Lenalidomide
In clinical trials or used based on other approved indications	TLK199 Src family kinase inhibitors Clotarabine	Arsenic trioxide Valproic acid Thalidomide	
Key supportive care concerns	Iron overload Cytopenias	Injection-site reactions GI toxicities	Fatigue

FAB = French-American-British Classification System; IPSS = International Prognostic Scoring System;

WHO = World Health Organization; WPSS = World Prognostic Staging System.

Adapted from Yarbro et al. *Cancer Nursing: Principles and Practice*. Jones & Bartlett Learning, Sudbury, MA; 2010.

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