

is critical to allow continuation of each treatment long enough to obtain optimal response (see Table 3). Early identification and prompt intervention for common adverse events will limit severity and reduce the probability of discontinuing treatment. Again, the majority of care is provided in the outpatient setting, with the patient and family bearing the bulk of the responsibility for early identification of adverse events. Patient and family education with consistent information, frequent reinforcement of key concepts, and active participation of the patient and family is critical to optimize outcomes.

Myelosuppression is the most common toxicity for all active therapies in MDS (Celgene Corporation, 2009, 2011; SuperGen Inc., 2010). Cytopenias often get worse before they get better, and patients may require continued transfusions before achieving hematologic improvement or transfusion independence. Given the median time to response in most patients of several weeks to months (Kurtin & Demakos, 2010; Silverman et al., 2011), these cytopenias may be disconcerting for the patient and the providers, who could view this as a sign of unacceptable toxicity or treatment failure. Setting expectations for

TABLE 4. MDS: Disease Snapshot

Feature	Key Findings
Epidemiology	15,000–20,000 new cases each year, with 35,000–50,000 existing cases. The average age at diagnosis is 72 years.
Etiology	Genetic instability, chemical exposure, tobacco use, mutagens, autoimmune disease, or simply unknown in the majority of cases (about 80%)
Stem cell defect Myeloid progenitor cell	Intrinsic factors (e.g., malignant clone, cytogenetic abnormalities) and epigenetic DNA modification (hypermethylation) Extrinsic factors (e.g., bone marrow microenvironment, stromal dysregulation, cytokine abnormalities) and imbalance of apoptosis and proliferation
Chromosomal findings Cytogenetic abnormality present in about 40% of cases	Favorable: -Y, del(5q), -20q Intermediate risk: +8 and other Poor risk: complex (more than three abnormalities); chromosome 7 abnormalities: 7q, -7, del(7p); inv16, t(8:12) indicative of acute myeloid leukemia
Additional prognostic factors indicating high-risk disease	Increased transfusion burden (more than two units in four weeks); increased blast cells (greater than 20% implies leukemic transformation); severe thrombocytopenia or neutropenia at diagnosis; atypical localization of immature precursors; bone marrow fibrosis, elevated ferritin, elevated lactate dehydrogenase—considered unfavorable; and 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 criteria 2006
Disease characteristics (all are incurable)	IPSS low and intermediate-1 risk: indolent course; low probability of leukemic transformation IPSS intermediate-2 and high risk: rapidly progressive course with early transformation to acute leukemia
Clinical presentation	Cytopenias (anemia most common), fatigue, infection, and bleeding
Treatment triggers	Transfusion dependence, progressive or symptomatic cytopenias, increased blasts
Key concepts for effective treatment	Supportive care alone does not prevent disease progression (no effect on the underlying disease). Disease-modifying therapies for MDS generally require a minimum of four to six months to achieve response; premature discontinuation may limit potential for an optimal response. Treatment should continue until disease progression or unacceptable toxicity. Aggressive concurrent management of cytopenias is essential to effective therapy. Treatment goals include reduced transfusion burden, delayed time to leukemic transformation, improved quality of life, and prolonged survival. Chromosomal abnormalities have prognostic value.
FDA-approved therapies	Azacitidine, decitabine, and lenalidomide
In clinical trials or used based on other approved indications	TLK199, src family kinase inhibitors, clofarabine, arsenic trioxide, valproic acid, and thalidomide
Key supportive care concerns	Iron overload, cytopenias, injection site reactions, gastrointestinal toxicities, fatigue, and rash (with lenalidomide)
FAB—French-American-British classification system; FDA—U.S. Food and Drug Administration; inv—inversion; IPSS—International Prognostic Scoring System; MDS—myelodysplastic syndromes; WHO—World Health Organization; WPSS—WHO Prognostic Staging System <i>Note.</i> From "Leukemia and Myelodysplastic Syndromes," by S. Kurtin (p. 1392). In C.H. Yarbro, D. Wujcik, and B.H. Gobel (Eds.), <i>Cancer Nursing: Principles and Practice</i> (7th ed.), 2011, Sudbury, MA: Jones and Bartlett. Copyright 2011 by Jones and Bartlett. Adapted with permission.	