Myelodysplastic Syndromes

What are Myelodysplastic Syndromes?	Myelodysplastic syndromes (MDS) comprise a diverse group of diseases that are characterized by impaired blood cell production in the bone marrow due to genetic aberrations in the blood stem cells. MDS has been recognized for more than 50 years and is also called preleukemia because of the high risk of progression to an aggressive blood tumor called acute myeloid leukemia (AML). AML is characterized by the rapid growth of abnormal white blood cells that accumulate in the bone marrow and interfere with the production of normal blood cells.[1] The median age of people diagnosed with MDS is between 60 and 75. Although MDS can affect people of any age, the onset of the disease before the age of 50 is rare. MDS is found worldwide and its characteristics are universally similar. Currently, only regional or local data on the number of people affected by the disease are available.
Causes and Classification	The causes for the genetic lesions in the genome of the stem cells that lead to the development of MDS are not known. In about 10 percent of the cases, MDS develops secondary to treatment with radiation therapy or chemotherapy for other diseases. Other risk factors that may be associated with MDS are smoking and occupational exposure to solvents or agricultural chemicals.[2] Over the last few decades, great efforts have been made to categorize MDS patients according to disease outcomes. In an initial attempt to classify the syndromes, a system was developed based on the changes in blood cells and bone marrow tissue and divided MDS into the following subtypes:[3,4] • Refractory anemia (RA) • Refractory anemia with excess of blasts (RAEB) • Refractory anemia with ringed sideroblasts (RARS) • Chronic myelomonocytic leukemia (CMML) • Atypical chronic myeloid leukemia (a-CML) • Myelodysplastic syndrome associated with del(5q) • "Unclassified" MDS The most commonly used classification system today is the revised international prognostic scoring system (IPSS-R). It considers the following disease characteristics that are the most powerful prognostic indicators. • The percentage of abnormal immature blood cells (blasts) in the blood • The number and appearance of chromosomes of the stem cells • The number and degree of blood cell reduction, evaluating

	hemoglobin, platelets and absolute neutrophil count[5] According to this classification, patients are categorized into the following prognostic subgroups based on both survival and progression to AML: • Very low • Low • Intermediate • High • Very high The WHO adapted prognostic scoring system (WPSS) represents a second classification method. Instead of determining the number and degree of blood cell reduction, this classification system takes the requirement of blood transfusions into account and groups patients into five risk categories.[6] • Very low risk • Low risk • Intermediate risk • High risk • Very high risk
Symptoms	The symptoms of MDS are often non-specific and can be related to a reduction in the number of blood cells such as: • Shortness of breath • Chest pain • Weakness or chronic tiredness • Pale skin • Easy bruising or bleeding • Increased susceptibility to infection Some patients have no symptoms, and in those cases, MDS often remains undiagnosed. A diagnosis of MDS can be made following a routine physical examination accompanied by a blood test called a complete blood count.

References

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